A SYNDROME OF EXTENSIVE PERIPAPILLARY MYELINATED NERVE FIBRES, HIGH IPSILATERAL MYOPIA AND REFRACTORY AMBYLOPIA.

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ABSTRACT: Myelinated retinal nerve fibers are rare developmental anomalies present in 0.5- 1% of all eyes. Occur due to myelination of ganglion cells beyond the lamina cribrosa during embryonic development. Reports of amblyopia associated with myelinated nerve fibers have been reported in the ophthalmologic literature. We report a case of 16 yr boy presenting with triad of peripapillary myelinated nerve fibre, u/l high myopia and amblyopia.

KEY WORDS: Myelinated nerve fibers, U/L high myopia, Amblyopia.

CASE REPORT: A 16 yr old male presented to our hospital with complaints of defective vision in right eye noticed since 2 months, with patient being unaware of visual defect and was detected during a school screening programme. Patient did not have any other significant ocular or systemic history.

On examination anterior segment examination of both eyes was normal with clear lens. Extra ocular movements were normal. Fundus examination of right eye showed clear media, disc was obscured with blurred margins, vessels showed tortuosity in inferior fundus, myelinated nerve fibres were seen around the disc and extending superiorly along the arcuate fibres and involving nasal fundus of about 3 disc diameter and whole of inferior fundus. Macula showed a absent foveal reflex with myelination involving inferior, temporal and nasal parts (Figure 1). Fundus examination of left eye was normal (Figure 2).Retinal examination of both parents and sister were done and were normal.

Best corrected visual acuity in right eye was CFCF at 2 mts and in left eye was 6/6. Colour vision in right eye could not be assessed due to low visual acuity and was normal in left eye. Intraocular pressure by applanation tonometry was 16 mm of Hg in the right eye, and 18 mm of Hg in the left eye. Visual fields in right eye could not be assessed due to low visual acuity and was normal in left eye.

Retinoscopy revealed -12.5 DS -2.50X170^0 RE and -0.50x180^0 in LE. Cycloplegic refraction revealed -10 DS -1.5X170^0 RE and -0.50X180^0 in LE. Full cycloplegic correction was given and patching of eye was advised.

FFA of right eye showed slight obscuration of retinal detail below the areas of myelination with tortuosity of vessels in inferior fundus and slight alteration in architecture of retinal capillary network below the areas of myelination. No abnormal vessels were noted (Figure3). FFA of left eye was normal.

Figure 1

Figure 2

Figure 3

DISSCUSION: Myelinated retinal nerve fibers are rare developmental anomalies present in 0.5- 1% of all eyes[1]. Studies disagree on whether or not there is a gender predilection[1]. Straatsma et al. studied both eyes from 3,968 consecutive autopsy cases and found 37 eyes from 32 patients with myelinated retinal nerve fibers. In this series, males and females were equally affected[2]. Myelination of the anterior visual system begins centrally at the lateral geniculate body at 5th month of embryonic development and proceeds distally to chiasm by 6–7 months, the retrobulbar optic nerve by 8 months, and the lamina cribrosa at term. In some cases it may continue after birth but does not proceed intraocularly[3].

The explanation for the occurrence of intraocular myelination is unknown. An error in process of myelination, or in the formation of the sclera, is hypothesized to be responsible for the myelin's reaching the retina[2].Oligodendrocytes responsible for myelination of the CNS, are not normally present in the human retina, but histological studies confirm their presence in some areas of myelinated nerve fibers, and their absence in other areas[2]. Based on this information, myelin would be expected to be found only over the optic nerve head. However, Williams argues that isolated patches of myelin can be explained because myelin can move through the retinal nerve fiber layer. It is able to "settle" in an area with low nerve fiber layer density and become visible[4].

Myelinated nerve fibers appear as white to gray-white areas of retina, obscuring underlying retinal detail. In most cases, they radiate peripherally from the optic disc, most commonly superior and inferior temporally, but can appear isolated in the posterior pole[1]. Ellis et al. divided myelination into three forms: type one affects one temporal arcade, type two affects both temporal arcades and type three is not contiguous with the disc[5]. Myelinated fibers are bilateral in 17–20% of cases and, clinically, they are discontinuous with the optic nerve head in 19%. Isolated patches of myelinated fibers are occasionally found in the peripheral retina[3].

Visual acuity is usually not affected in these cases; however Patient may have relative scotomas if sufficient number of myelinated nerve fibres are present. Walsh and Hoyt observed a pronounced defect in visual acuity as a result of myelinated fibers. In that case, the entire fundus except for a small area temporal to the disc contained myelinated nerve fibers[3].

Studies have found a relationship between myelinated nerve fibers, amblyopia and high myopia with or without strabismus. Kodama et al[1]found that only 0.03% of patients with myelinated

nerve fibers had amblyopia and myopia while Straatsma et al[2] found that 10% of patients with myelinated nerve fibers had myopia, amblyopia and strabismus[6].

Ellis et al. found that 83% of patients with myelinated nerve fibers had myopia greater than 6 diopters[5].The authors postulated that medullated nerve fibers may contribute to myopia by creating axial enlargement. In contrast, other authors argue that the greater axial length of myopic eyes puts them at a greater risk for damage secondary to myelination[7].

Extensive unilateral myelination of nerve fibers can be associated with high myopia and severe amblyopia which unlike other forms of ambylopia is notoriously refractory to treatment[5]. In such patients, myelination surrounds most or all of the circumference of the disc. Additionally, the macular region (although unmyelinated) usually appears abnormal, showing a dulled reflex or pigment dispersion. Hittner et al. found the appearance of the macula to be the best direct correlate of response to occlusion therapy[9].

Authors cannot agree on the etiology of the amblyopia in patients with myelinated nerve fibers. Some authors assume there is a organic cause to the amblyopia. Williams proposes that the retinal myelination causes a decrease in the number of ganglion cells which causes optic nerve hypoplasia, resulting in decreased visual acuity[4].Holland and Anderson, postulated that the myelinated nerve fibers result in an elevation of the optic disc which may lead to an overall disorganization of neural elements. This disorganization of visual pathways is responsible for amblyopia development[8].

Rarely, areas of myelinated nerve fibers may be acquired after infancy and even in adulthood. Trauma to the eye (a blow to the eye in one patient and an optic nerve sheath fenestration in the other) seems to be a common denominator in these cases[10] . Williams suggested that sufficient damage to the lamina cribrosa may permit oligodendrocytes to enter the retina, whereupon they move to the nearest area of fairly loose nerve fibers and myelinate them[4]. Conversely,myelinated nerve fibers disappear as a result of tabetic optic atrophy, pituitary tumor, glaucoma[11], central retinal artery occlusion, branch retinal artery occlusion[12], and various optic neuropathies.

CONCLUSION: In our case study there are two factors that can contribute to amblyopia U/L high myopia and myelinated nerve fibres. However it's difficult to differentiate which is the primary and secondary factor. In some studies patients had abnormal macular areas, which could also be responsible for the decrease in vision and in some the amblyopia could be attributed to high refractive error. This triad of U/L high myopia, peripapillary myelinated nerve fibre and amblyopia has been presented for its rarity.

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