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Straatsma syndrome: Unilateral myelinated retinal nerve fiber layer, high myopia, strabismus and amblyopia: Case report

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Abstract
Straatsma syndrome is a rare disease entity characterized by the traditional triad of unilateral myelinated retinal nerve fibers (MNRF), axial myopia, and amblyopia. The original description of the condition also included strabismus· Variations of the triad include a "reverse Straatsma syndrome", in which patient's exhibit hyperopic instead of myopia. Even though nystagmus and strabismus have not been prominently associated with Straatsma syndrome, either may be present as complimentary findings without precluding one from the diagnosis. A 12-year-old boy presented with report of decreased vision in the right eye for the last year. On examination, his best-corrected visual acuity was 1.78 log MAR in the right eye and 0 log MAR in the left eye. Cycloplegic retinoscopy revealed a refractive error of a −15 dioptre sphere with a −3.5 dioptre cylinder at 180° in the right eye. The patient had right exotropia of 10 prism diopters on modified Krimsky's test with full extra ocular motility.

Keywords: Straatsma syndrome, unilateral myelinated retinal nerve fibers, strabismus, amblyopia

Introduction
Straatsma syndrome is a rare disease entity characterized by the traditional triad of unilateral myelinated retinal nerve fibers (MNRF), axial myopia, and amblyopia. The original description of the condition also included strabismus. Variations of the triad include a "reverse Straatsma syndrome", in which patients exhibit hyperopia instead of myopia.

Case Description
A 12-year-old boy presented with report of decreased vision in the right eye for the last year. On examination, his best-corrected visual acuity was 1.78 logMAR in the right eye and 0 logMAR in the left eye. Cycloplegic retinoscopy revealed a refractive error of a −15 dioptre sphere with a −3.5 dioptre cylinder at 180° in the right eye. The patient had right exotropia of 10 prism diopters on modified Krimsky's test with full extraocular motility. Dilated fundus examination was performed, which showed the presence of a myelinated retinal nerve fiber layer in the right eye along the superior and inferior temporal arcades covering the whole posterior pole with a dull foveal reflex (figure 1A). The left eye fundus was normal (figure 1B). The axial length of the right eye was 27.60mm and that of the left eye was 22.07mm. Optical coherence tomography was performed, which depicted parafoveal hyper-reflective retinal nerve fiber layer in the right eye and left eye was normal (figure 2). An optical correction was prescribed in the form of a contact lens in the right eye and occlusion therapy was given in the left eye.

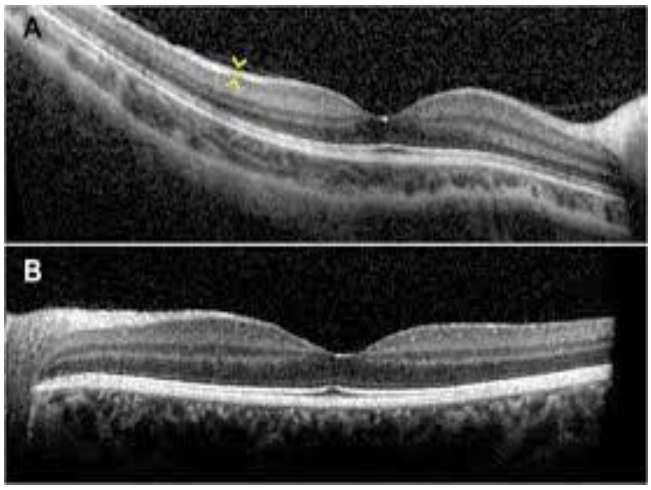

Fig 1: Dilated fundus examination was performed, which showed the presence of a myelinated retinal nerve fiber layer in the right eye along the superior and inferior temporal arcades covering the whole posterior pole with a dull foveal reflex (figure A). The left eye fundus was normal (figure B)

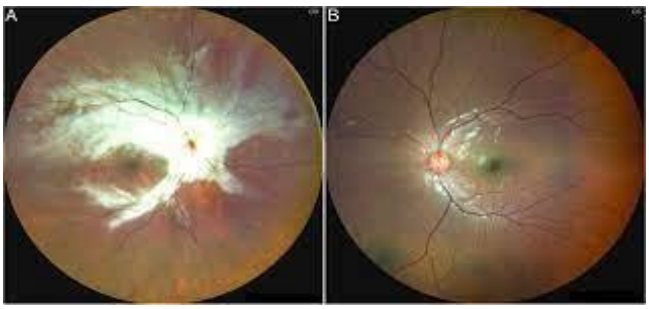

Fig 2: (A) Fundus photo of the right eye showing extensive myelination of the retinal nerve fiber layer. (B) Normal fundus of the left eye.

Discussion

Straatsma syndrome is defined as myelinated nerve fiber with myopia and amblyopia. Myelination of the optic nerve begins from the lamina cribrosa and the lamina cribrosa acts as a barrier at term. Defects in the lamina cribrosa or ectopic oligodendrocyte progenitor cells lead to the myelinated retinal nerve fiber layer. An optical correction was prescribed in the form of a contact lens in the right eye and occlusion therapy was given in the left eye. Amblyopia, if detected early in life, can be treated with good visual outcomes.

Conclusion

I would like to state that patients started with a chief complaint of decreased vision in the right eye for the last year. He was treated with an optical correction was prescribed in the form of a contact lens in the right eye and occlusion therapy was given in the left eye. There was no improvement in the visual acuity after 6 months of amblyopia treatment. Amblyopia, if detected early in life, can be treated with good visual outcomes.

Consent for publication

Informed consent was obtained from parents of the patients to publish this case in medical journal.

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