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A rare case of Straatsma syndrome

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Abstract
The presence of retinal myelinated nerve fibers is not a rare finding during routine examinations, and it is usually a benign and isolated finding. However, in some rare cases, it can be associated with other ophthalmological conditions. We describe a case of a patient with the triad of myelinated nerve fibers, myopia and amblyopia, which configures the Straatsma Syndrome.The originaldescription of the condition also included strabismus.Variations of the triad include a "reverse Straatsma syndrome", in which patient's exhibit hyperopia 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.
Patients are usually asymptomatic, but a few cases can present with significant visual abnormalities, commonly axial myopia and amblyopia, configuring the Straatsma Syndrome. We report a case of a 28 years old female patient with this rare syndrome.
A 28-year-old female presented with complains of decreased vision in the left eye since childhood . On examination, her best-corrected visual acuity was 6/6 ,N6 in the right eye and counting finger 3 meter,N36 in the left eye. Dry refraction revealed a refractive error of +0.25 dioptre sphere with a −0.25 dioptre cylinder at 90° in the right eye and -7.00 diopter sphere in the left eye. The patient had left exotropia of 30 prism diopters on modified Krimsky's test with full extra ocular motility.

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

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I. Introduction

Straatsma syndrome was originally described by Straatsma et al.[1] in 1979 in a case series of 4 patients with unilateral myopia, amblyopia, and strabismus associated with myelinated retinal nerve fibers (MRNF). With the growing literature, the triad of MRNF, myopia, and amblyopia is now accepted as Straatsma syndrome.[2] However, additional findings such as strabismus, nystagmus, hypoplastic optic nerve, and heterochromia iridum have also been reported and do not preclude the diagnosis of this syndrome.[2,3,4] There is even a reported variation of the triad with hyperopia instead of myopia, called "reverse Straatsma syndrome."[5] Although it is generally unilateral, bilateral cases of traditional and reverse Straatsma syndrome have also been reported.[2,6]

The challenging part of the syndrome is treating amblyopia. Several factors are reported to be associated with poor visual outcomes after occlusion therapy, including a high degree of anisometropia, strabismus, extensive myelination, and macular involvement.[3,7,8,9]

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II. Case Description

A 28-ye111ar-old female presented with the complaint of decreased vision in the right eye since childhood. On examination, her best-corrected visual acuity was 6/6 ,N6 in the right eye and counting finger 3 meter ,N36 in the left eye. The patient had left exotropia of 30 prism diopters on modified Krimsky's test with full extraocular motility. Dilated fundus examination was performed, which showed the right eye fundus has normal disc findings and dull foveal reflex (figure 1A) and presence of a myelinated retinal nerve fiber layer in the left eye along the superior and inferior temporal arcades covering the whole posterior pole with a dull foveal reflex (figure 1B). The axial length of the right eye was 23.84 mm and that of the left eye was 27.56 mm (IOL master 500). SD OCT (Spectral domain optical coherence tomography, Topcon) was performed, which depicted

parafoveal hyper-reflective retinal nerve fiber layer in the left eye(figure 2B) and no macular or optic disc alterations were found in the right eye (figure 2A).

III. Discussion

The pathogenesis of myelinated nerve fiber is not completely known. It is believed that can result from an imbalance between the formation of the lamina cribrosa, which proceeds posteriorly from the limbus and the process of myelination ,which begins from the lateral geniculate body[10].Oligodendrocytes are effectors in the myelination of the ganglion cell axons and impaired astrocyte function causes those cells to migrate through lamina cribrosa ,is taught to play a role [11].

Myopia has been reported in 35%to 58% in patients with myelination of fibers in previous studies,83% with refraction of up to 6 diopters[12].One hypothesis for this association is that a blurred image on the retina produces visual depravity at a critical stage of ocular development, which could be the cause of axial elongation of the eye and development of of myopia, which in turn delays the development of the lamina cribrosa, thus following an extension of myelination of fibers through the retina[10].

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. [10]

In this patient, an optical correction was prescribed in the form of a contact lens in the left eye and occlusion therapy was given in the right eye. Six monthly follow up had been advised.

The prognosis of this entity is poor to moderate, particularly if the condition is not diagnosed during the amblyogenic period, giving rise to a picture of severe amblyopia.Due to its rarity and features not widely known ,straastsma very often goes undiagnosed.

IV. Conclusion

I would like to state that patients started with a chief complaint of decreased vision in the left eye since childhood. She was treated with an optical correction.It was prescribed in the form of a contact lens in the left eye and occlusion therapy was given in the right eye.The patient had been advised 6 monthly follow up.
Amblyopia, if detected early in life, can be treated with good visual outcomes.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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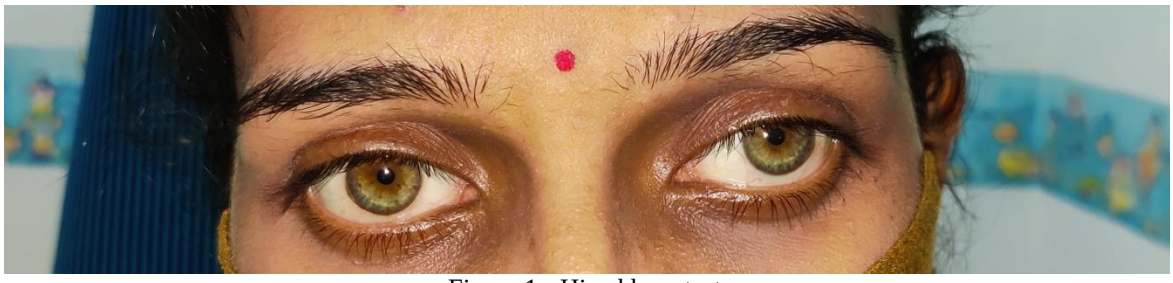

Figure 1 - Hirschberg test

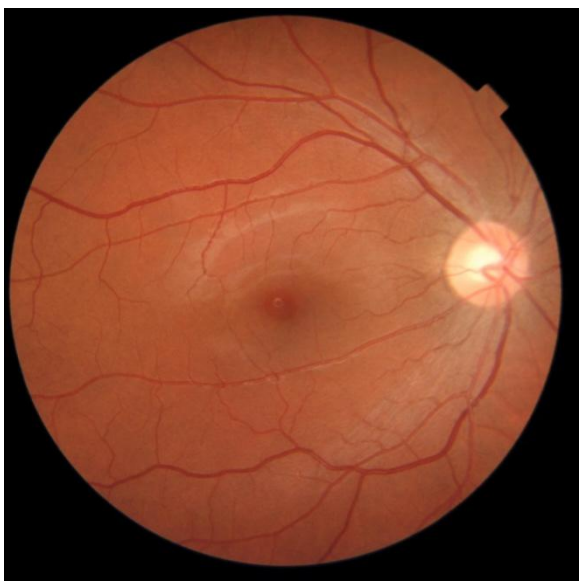

Figure 1A -Wide field fundus photography Of right eye

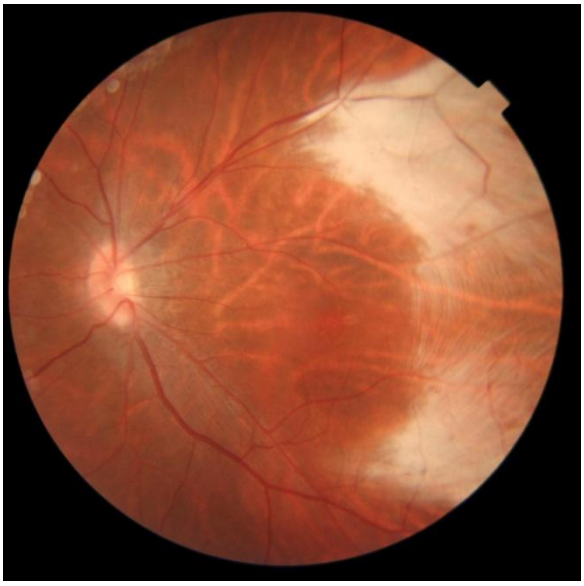

Figure 1B- Wide field fundus photography of left eye

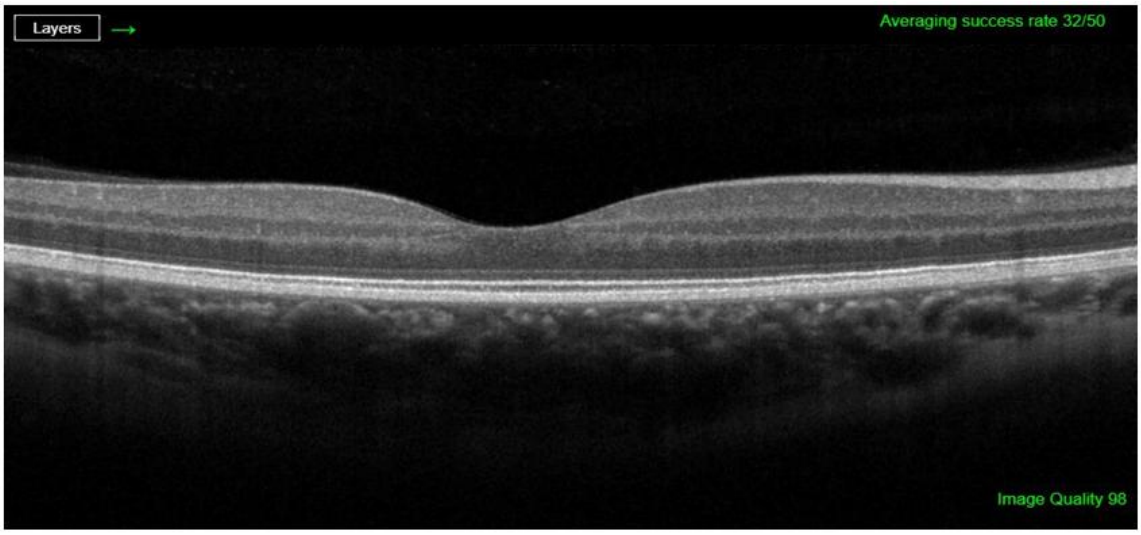

Figure 2A- Foveal centered SD OCT scan of right eye

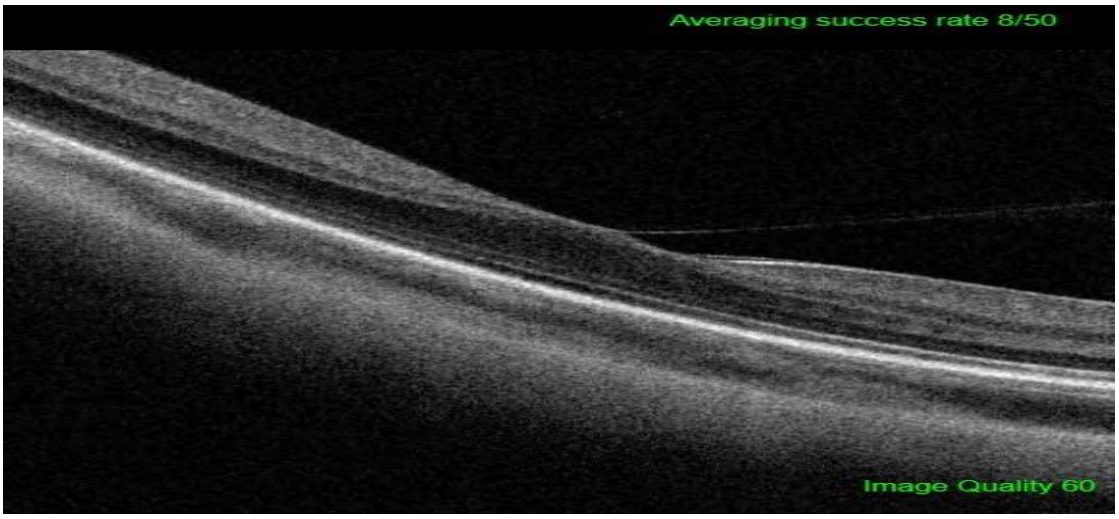

Figure 2B- Foveal centered SD OCT scan of left eye

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