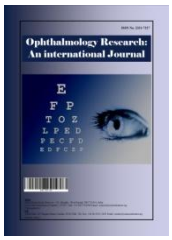

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Reverse Straatsma Syndrome – A Case Report

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Objective: Ophthalmologic signs and symptoms were evaluated in a patient diagnosed with reverse Straatsma syndrome.
Case Presentation: 22 years old male presented to Eye OPD with unilateral diminution of vision of left eye present since childhood. Complete ophthalmologic examination showed anisometropic hypermetropic ambylopia of +8DS/ +3.5DC x 90◦ in LE, correctable to 6/60 and myelinated nerve fibres in LE retina superior to optic disc. The RE after refractive correction of +1.5 DS showed 6/6 vision.
Results: Patient was given full refractive correction of RE and counseled for the condition in LE.

Keywords: Reverse straatsma syndrome; hypermetropia; amblyopia; myelinated nerve fibres; refractive correction.

1. INTRODUCTION

First described by Virchow in 1856, [1] very often, myelinated nerve fibres are isolated findings which are incidentally detected on detailed examination of retina appearing as whitish- yellow network like thin membrane and obscuring the view of underlying retinal structures and they are very rarely associated with diminution or loss of vision. Sometimes, it is associated with refractive errors (myopia, hypermetropia), amblyopia or strabismus. G. Dimitrova et al. published a case series in which they studied a variety of anatomical and

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clinical manifestations of myelinated nerve fibers [2].

In a series of 3,968 consecutive autopsy cases, Bradley R Straatsma and colleagues reported that myelinated nerve fibres were present in 0.98% of patients and in 0.54% of eyes examined, with bilateral involvement in 7.7% of patients [3]. Thus, Bradley R Straatsma et al. [4] first described Straatsma syndrome in 1979 in a case series of 4 patients. Straatsma Syndrome is a rare condition which presents as significant diminution of vision associated with a triad of amblyopia, myopia and myelinated nerve fibres in retina [1], sometimes associated with strabismus [4].

Reverse Straatsma Syndrome is a variation in this triad, which is even rarer and exhibits unilateral or bilaterally asymmetrical hypermetropia instead of myopia, amblyopia and myelinated nerve fibres in retina [1]. Here, we document a rare case of a young patient of Reverse Straastma Syndrome.

1.1 Aims and Objectives

To evaluate ophthalmologic signs and symptoms in a patient diagnosed with reverse Straatsma syndrome.

2. CASE PRESENTATION

A 22 years old male patient from lower-middle class, working as a shopkeeper, presented to eye OPD with unilateral diminution of vision of left eye (LE) which was present since childhood but he never paid due attention and ignored it. Complete and detailed ophthalmological examination was carried out in his both eyes. Preliminary examination showed –unaided visual acuity (VA) of counting fingers at 1m (CF@1 m) in LE and 6/9(P) in RE. Refraction was done in both eyes to find out BCVA and amount and type of refractive error. Eyes were examined in primary position to check for strabismus and extra-ocular movements were checked in all gazes to note any restricted movement and conjugate/disconjugate movements. Complete anterior segment (AS) examination was done with torch light (pupillary reflexes) and Slit Lamp Biomicroscope. Intraocular pressure (IOP) was recorded in both eyes with Goldmann Applanation Tonometry. Dilated fundoscopy of both eyes were done with indirect ophthalmoscopy and fundus pictures were taken with fundus camera. A-scan biometry was done in both eyes to record their axial length. Complete systemic examination was done to rule out tumors, inflammation etc.

3. RESULTS

Best Corrected Visual Acuity (BCVA) in right eye (RE) was 6/6 with refraction (+1.5DS) and left eye (LE) was 6/60 with refraction (+8DS/ +3.5DC x 90◦). Both eyes were orthotropic (aligned straight) and there was complete range of movement and conjugate movement of both eyeballs in all gazes and positions. On torch light examination, pupils of both eyes (BE) were normal in size and normal light reflexes (direct and consensual) were present. Slit lamp biomicroscopy revealed normal structures in Anterior Segment of both eyes (BE). Intraocular Pressure (IOP) recorded by Goldmann Applanation Tonometer (GAT) measured 13 mmHg in left eye (LE) and 12mmHg in right eye (RE). Indirect Ophthalmoscopy with a +20D lens showed normal fundus in right eye (RE) but left eye (LE) fundus showed myelinated nerve fibres, emerging from optic disc and present superior, inferior and temporal to optic disc. Nasal retina and macula were free from myelination. Macula was normal (Foveal reflex seen). Optic disc showed a cup-disc ratio (C:D) of 0.4. A-Scan biometry done by immersion technique revealed axial length of: LE – 20 mm and RE – 23 mm.

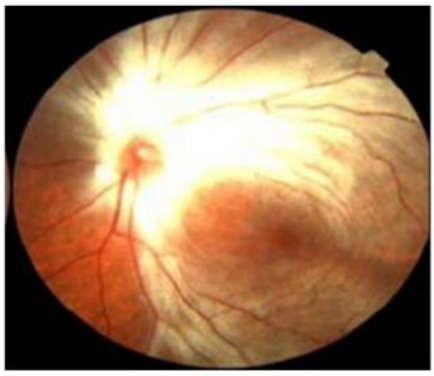
4. DISCUSSION

Literatures quote that normally during embryonic development, nerve fibres entering the retina lose their myelin sheath at ora serrata, thus retinal nerve fibres are unmyelinated. The myelination process normally terminates at the level of lamina cribrosa at about 8^{th} month of gestation till birth, but occasionally it continues into the retinal nerve fiber layer, giving it a whitish – yellow net-like appearance. Myelinated retinal nerve fibres result from the faulty location of oligodendrocyte-like cells in retina, prior to development or temporary loss or absence of the barrier function of the lamina cribrosa at optic nerve head [5].

Three different types of myelination are known [6]-

- type 1 pattern along the superior temporal arcade
- type 2 pattern along both the arcades
- type 3 pattern with no contiguity with the ONH.

Right eye – normal fundus



Left eye – mvelinated nerve fibres in superior, inferior and temporal quadrant
Macula - normal

Fig. 1. Showing myelinated nerve fibres in superior, inferior and temporal quadrants of retina, emerging from optic nerve head in left eye

Our patient had type 2 myelination.

Myelinated retinal nerve fibres is a congenital and stationary condition which is more common in myopic than in hyperopic eyes, but there are some reports of myelinated fibers associated with hyperopia, which may be unilateral or bilateral [7,8].

There are very few cases of Straatsma Syndrome and Reverse Straatsma Syndrome (<50) reported in literature making it rare and not much information is available on the pathophysiology of occurance and it is unclear that whether diminution of vision is due to myelinated nerve fibres or a consequence of anisometropia. A case report of bilateral Reverse Straatsma Syndrome published by Shenoy R et al. [9] postulated the etiology being anomalous distribution of oligodendrocytes in retina thus leading to myelination of retinal nerve fibres.

In our case, anisometropic amblyopia seems to have a stronger influence on the relative visual acuity of the patient's eyes than the presence of retinal nerve fiber myelination which is supported by Kee and Hwang [10] that prognostic factors for the visual improvement in amblyopia were the amount of anisometropia and the area of myelination. With time, he developed anisometropic amblyopia and myelinated nerve fibres turned out to be an incidental finding.

Generally, such a large amount of anisometropia (more than +5D in our case) leads to development of squint in early age itself, but strangely our case had straight aligned eyes and no phorias were also noted. Some cases reported in literature have strabismus associated with them.

5. CONCLUSION

Our patient was given full refractive correction of right eye (+1.50DS) and left eye was left unaided and he was counseled for the condition and visual prognosis in left eye.

Visual prognosis of amblyopia associated with myelination of retinal nerve fibers and anisometropia is poorer than anisometropic amblyopia without myelination. It is refractory to occlusive therapy. Despite having a poor prognosis, visual rehabilitation should be attempted.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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