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Case Report

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Reverse straatsma syndrome and lamellar cataract: a case report and review of the literature

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ABSTRACT

Myelinated retinal nerve fiber (MRNF) is a developmental anomaly that is often detected during routine ophthalmological examination, but can be an ocular finding in a wide variety of diseases. An important example of this group is the Straatsma Syndrome. Straatsma Syndrome is an entity that presents with the triad of myopia, amblyopia, and MRNF. When this triad is accompanied by hyperopia instead of myopia, it is referred to as the Reverse Straatsma Syndrome. Here, we present a case of Reverse Straatsma Syndrome accompanied by lamellar congenital cataracts.

Keywords: Amblyopia, hypermetropia, myelinated retinal nerve fiber, reverse straatsma syndrome

INTRODUCTION

Myelinated retinal nerve fiber (MRNF) is a developmental anomaly with blurry borders located in the retinal nerve fiber layer and often has a white/gray-white appearance. It was first described by Virchow in 1856.[1,2] Although it is a finding frequently encountered during routine ophthalmological examination, it can also appear as a symptom of a wide variety of diseases (Gorlin syndrome, neurofibromatosis, etc.).[2] Straatsma Syndrome is the name given to the myopia and amblyopia triad accompanying MRNF. There is also a variation called Reverse Straatsma Syndrome, which may present with a hyperopia triad instead of myopia.[3] This article aims to present and discuss a case of unilateral Reverse Straatsma Syndrome accompanied by lamellar cataract.

CASE

A 26-year-old male patient was admitted to our retina unit with a complaint of decreased vision in his left eye, which had been present since childhood. There was no additional systemic disease in the patient's medical history. The patient was diagnosed with amblyopia and mild cataracts in his previous examinations. On visual acuity examination, the best-corrected visual acuity (BCVA) of the right eye was 20/20, and the left eye was counting fingers at 5 meters (with +14 D spherical correction). Intraocular pressure was normotonic bilaterally. The anterior segment examination revealed a normal appearance in the right eye and a lamellar cataract in the left eye. The visual axis was open (Figure 1). Dilated fundus examination revealed that the right eye was normal. In the left eye, there was an MRNF image with blurry borders, feathery, and white, shading the retinal vessels that started from the disc and spread 360°. No foveal involvement was observed (Figure 2). Based on these findings, the patient was diagnosed with Reverse Straatsma Syndrome. The patient's follow-up continued in our retinal unit.

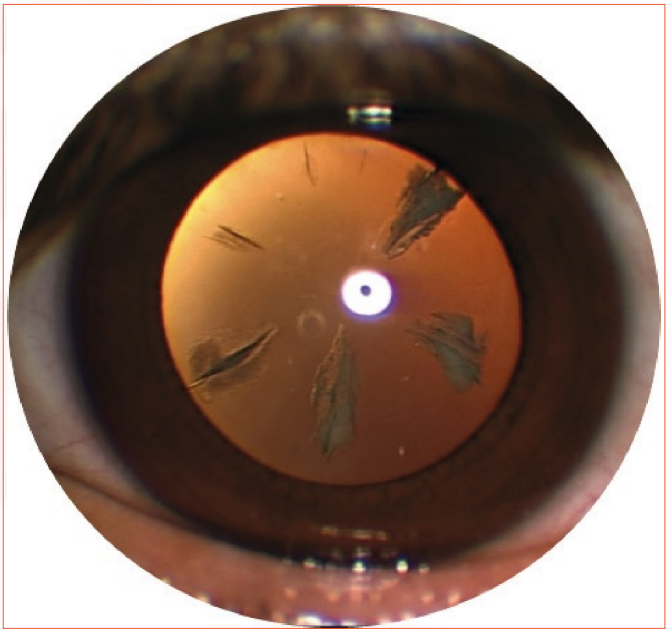

Figure 1. Lamellar cataract appearance observed in the anterior segment photograph of the patient's left eye.

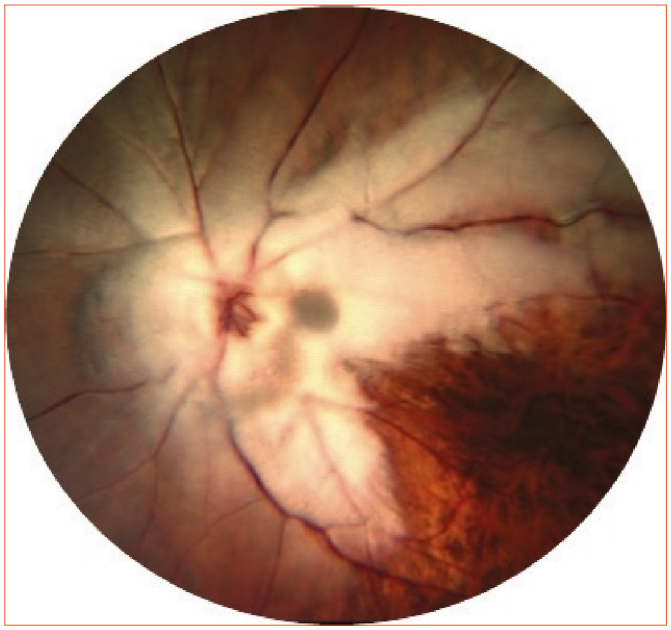

Figure 2. In the colored fundus photograph of the patient's left eye, an image of a feathery myelinated nerve with faint borders spreading 360° from the disc is observed.

DISCUSSION

In an autopsy study by Straatsma et al.[1] who examined 7936 eyes of 3968 patients, they observed that 39 of the patients (0.98%) had MRNF, and three of these patients (7.7%) presented bilaterally. In total, MRNF was detected in 42 eyes (0.54%), and there was no statistically significant difference in terms of sex between patients. The pathogenesis of MRNF is still not fully understood. According to one opinion, the abnormal distribution of oligodendrocytes is held responsible for the development of this condition.[1,4] Another thought is that the blurring of the retinal image during ocular development causes visual stimuli to decrease, resulting in axial elongation of the eye and the development of myopia. During this axial elongation, the development of the lamina cribrosa is disrupted, and it is thought that it cannot function as a sufficiently strong barrier to prevent the migration of oligodendrocytes to the retina.[2] This idea contradicts cases with Reverse Straatsma syndrome, such as in our case, and supports the idea of abnormal distribution of oligodendrocytes as the pathogenesis.[1,4,5]

It has been stated that the presence of wide MRNF may be associated with high myopia and poor visual acuity. In fact, in a study evaluating 12 patients with MRNF, poor visual acuity after treatment was associated with the myelination area around the fovea, and retinal involvement of nine clock dials and more had the worst visual outcome.[6] In the study conducted by Hittner et al.[7] it was determined that the appearance of the macula is an important factor in predicting visual acuity in the presence of MRNF and anisometropia. They also found that eyes that responded poorly to amblyopia treatment had abnormal macula. In the study conducted by Sevik et al.[8] the presence of profound anisometropia, strabismus myelination, and macular involvement were expressed as poor prognostic features and were thought to be associated with poor visual outcomes after treatment. In the case of bilateral Reverse Straatsma Syndrome presented by Alenezi et al.[5] myelinated nerve fibers were located peripapillary in both eyes and the macula was observed naturally. The authors attributed the low visual acuity in this case to ametropia and stated that this situation was more important than the presence of MRNF. We think that the low visual acuity in our case was mostly due to the presence of anisometropia. Although myelinated nerve fibers are located in the macula, the relative preservation of the fovea was an important factor that led us to think that the reason for low visual acuity was anisometropia. A wide variety of ocular findings accompanying the triad of myopia, amblyopia, and MRNF has been described in the literature in cases of Straatsma Syndrome. Examples of these ocular findings include nystagmus, heterochromia iridis, strabismus, and optic nerve hypoplasia.[2,8,9] In addition, a recently published case report stated that this picture may be accompanied by congenital cataracts and that this association may have occurred with common etiopathogenic mechanisms.[10] To our knowledge, this is the first report of a case of congenital cataract accompanying Reverse Straatsma Syndrome. This association between Reverse Straatsma Syndrome and congenital cataract may be related to a common developmental etiopathogenesis, or it may be related to the simultaneous occurrence of two different entities.

CONCLUSION

As a result, Reverse Straatsma Syndrome is a rare entity that can present with varying degrees of visual acuity decrease, depending on the MRNF width and anisometropia depth. These cases may occur simultaneously with congenital cataracts and Reverse Straatsma Syndrome.

ETHICAL DECLARATIONS

Informed Consent

All patients signed and free and informed consent form.

Reviewer Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors declare no potential conflicts of interest.

Financial Disclosure

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Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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