American Journal of Ophthalmology Case Reports 8 (2017) 71–73

Contents lists available at ScienceDirect

American Journal of Ophthalmology Case Reports

journal homepage: http://www.ajocasereports.com/

Case report

Unusual Straatsma Syndrome - How dogmatic is a bad prognosis?

Ana Vide-Escada [a, *], Helena Prior Filipe [b]

[a] Integrated Responsibility Centre of Ophthalmology of Hospital Garcia de Orta EPE, Av. Torrado da Silva, 2805-267 Almada, Portugal
[b] Hospital das Forças Armadas/Polo de Lisboa-EMGFA, Azinhaga dos Ulmeiros, 1690-020 Lisboa, Portugal

ARTICLE INFO

Article history:
Received 26 April 2016
Received in revised form
7 April 2017
Accepted 2 October 2017
Available online 7 October 2017

Keywords:
Heterochromia *iridum*
High myopia
Anisometropia
Amblyopia
Myelinated retinal fibres
Esotropia

ABSTRACT

Purpose: To show that Straatsma Syndrome can have a good outcome and to highlight an unusual presentation of this disease.
Observations: A four-year-old boy presents with severe right eye amblyopia in association with high myopia, esotropia, heterochromia *iridum* and extensive myelinated retinal fibers involving both temporal arcades and the optic nerve head. Right eye initial visual acuity was less than 20/400 for distance and less than R6W10 for near. Left eye examination was unremarkable. Despite the indicators for bad prognosis, intensive occlusion therapy was prescribed. Parents were strongly involved in the treatment regimen. After four months, the patient presented an unexpected good visual recovery both for distance and near, that has persisted until present. Right eye visual acuity is 20/30 with −9.00 dioptres contact lens for distance and R2W1 for near. Esotropia also improved to 12 prism dioptres. Fundoscopic alterations and heterochromia *iridum* have remained stable. Spectral-domain optical coherence tomography images of the right eye showed thinner superior outer ring measurements.
Conclusions and importance: Straatsma Syndrome can present with heterochromia *iridum*. When strabismus is present, early surgery should be withheld. Intensive treatment of Straatsma Syndrome can yield an unexpected good result, despite initial high degree anisometropia and low vision acuity.

1. Introduction

Myellinated retinal nerve fibers are present in about 1% of patients and were first described by Virchow.[1,2] The cause for this abnormality is still not fully understood. One theory states it might be related to the failure of the lamina cribosa in detaining the anterograde optic nerve myelination, while another points out the presence of heterotopic oligodendrocytes-like cells as the *primus movil*.[3,4]

Straatsma described a specific syndrome comprising amblyopia, high myopia and myelinated retinal fibers (MRF).[5] Strabismus is a frequent finding, in opposition to heterochromia *iridum*. It is often isolated but can be associated with retinal vascular, retinal membrane, ocular developmental and craniocephalic abnormalities, as well as hamartoneoplastic and familiar disorders.[3] Prognosis in uncertain as amblyopia is multifactorial.

Recently, optical coherence tomography (OCT) analysis in these patients have been performed.[6,7] Central macular thickness is reported to be significantly thicker and the outer ring macular thickness significantly thinner.[8] In Straatsma Syndrome (STAS), the latter is thought to be unrelated to the degree of myopia, but rather due to the shadowing of the outer layers of the retina induced by the medullated nerve fibers, causing imprecise OCT measurements.[6]

The authors report a case of STAS, strabismus and heterochromia *iridum* documented by SD (Spectral-domain) OCT presenting an unexpected good outcome.

2. Case report

A four-year-old boy presents with severe amblyopia, high myopia, photophobia, heterochromia *iridum* and right esotropia(Fig. 1). He is a healthy dizygotic twin, with no personal or family relevant history.

At presentation, right eye (RE) visual acuity (VA) was less than 20/400 for distance and less than R6W10 (Rossano Weiss – RW) for near. Prior refraction data was −7.00−0.50 85° RE and plano left eye (LE). A 30 prism dioptre (PD) right eye esotropia was found. Fundoscopy revealed extensive MRF involving both temporal arcades and contiguous with the optic nerve head (ONH) (Fig. 2). Fixation

* Corresponding author.
E-mail addresses: anavideescada@gmail.com (A. Vide-Escada), hpriorfilipe@gmail.com (H. Prior Filipe).

https://doi.org/10.1016/j.ajoc.2017.10.003

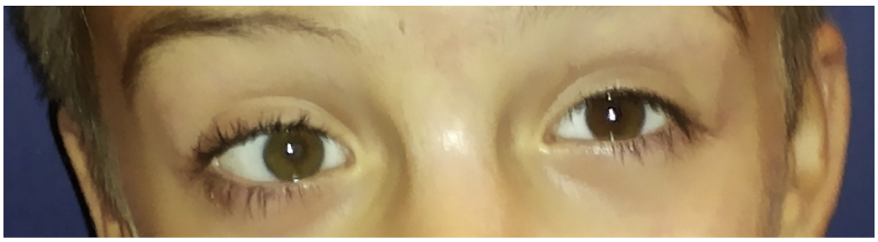

Fig. 1. Heterochromia *iridum*.

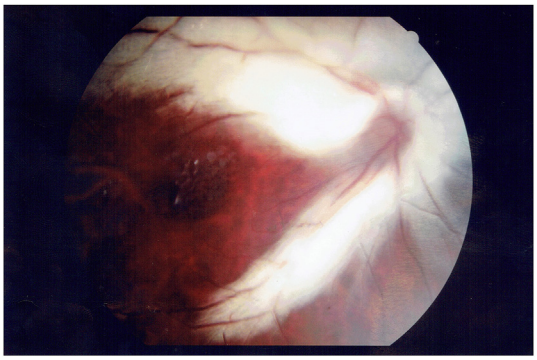

Fig. 2. Fundoscopic appearance of the right eye.

was central, although not foveal, and unstable. LE examination was unremarkable and LEVA was 20/20.

Despite all bad prognostic signs, a permanent three-week occlusion of the LE was prescribed and tolerated by the child. Parents were actively engaged in the treatment plan since the beginning.

A slight improvement was documented after this period: REVA 20/250 for distance and R4W6 for near. Therefore, an intensive full-day patching regimen of 1/6 (one day RE occlusion and six days LE occlusion) was then prescribed and kept for nine months when the boy started school. Since then, a 5 hours/day LE occlusion regimen has been followed.

A year and a month following the initial presentation, a silicon-hydrogel contact lens (CL) of −9.00 dioptries (D) was fit into the RE, to which the patient adapted well and has been wearing ever since.

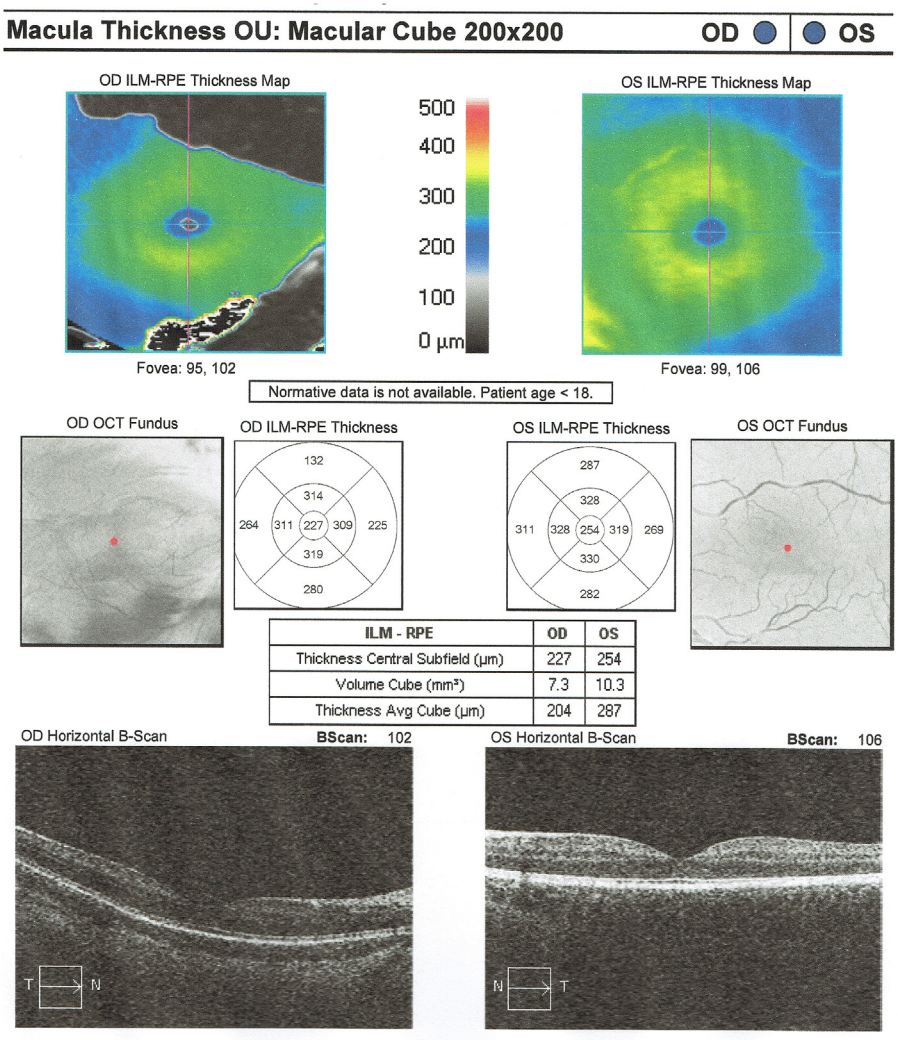

Fig. 3. Spectral domain optical coherence tomography images of the right eye showing normal foveal contour and thinner superior outer ring measurements.

SD OCT images of the RE show normal foveal contour and thinner superior outer ring measurements (Fig. 3).

Last consultation revealed REVA 20/30 for distance and R2W1 for near, right eye esotropia improvement to an aesthetically acceptable 12 PD, despite absence of binocular vision and stable heterochromia *iridum*. Fundus alterations have remained stable.

3. Discussion

This is a case report of STAS with an unexpected good outcome. The type of fixation, the age of diagnosis, the initial VA, the extensive area of myelinisation, the high myopia and the high anisometropia all indicated a most likely bad prognosis. Moreover, the extreme difficulty in tolerating an occlusion therapeutic regimen was an additional challenge.

Fundoscopy was fundamental for diagnosis and is a mandatory step in the evaluation of every child. Additional important information is also retrieved by evaluating the type of fixation.

In our case, type two MRF was present, less common than isolated myelin islands and usually associated with a worse prognosis.[2,9] An unusual association with STAS was found – heterochromia *iridum*.

Associations between final VA and the degree of anisometropia and the presence of strabismus have been reported.[3] One paper reports VA of 20/30 or better with mean anisometropia of −6.38 D, but several other show disappointing results.[10–12]

Classically, STAS is associated with axial myopia instead of refractive, thus apparently excluding CL wear benefit.[5] However, in our case, CL wear was successful and should therefore be tried.

The amblyopia resulting from myopia and MRF is dual: high myopia might itself induce defocus and MRF might interfere directly with vision.[5] In spite of this, VA has successfully improved, probably due to the foveal region's sparing, as was shown by OCT, despite the presence of photophobia and the type of fixation.[13] The RE superior outer macular ring was thinner, which is similar to other reports.[6,14]

The amblyopia was so severe that a full-day patching occlusion regimen was instituted. We hypothesized this contributed undoubtedly to the success achieved since it allowed a brain “switch on” of the RE visual pathway and also had a stabilizing effect in the type of fixation. This last mechanism probably explains why esotropia improved to 12 PD without any specific treatment.

4. Conclusions

This case report clearly demonstrates the importance of early parents engaging and emphasises the fact that in children it is mandatory to try to improve vision despite all bad prognostic indicators. The improvement of vision can by itself lead to an improvement in strabismus and so early strabismus treatment should be withheld. In addition, this case reveals an unusual association between STAS and heterochromia *iridum*.

5. Patient consent

Parents were informed and engaged actively in the treatment plan and gave oral consent to using the child's data for scientific work.

Funding

No funding or grant support.

Conflict of interest

The authors have no financial disclosures relating this topic.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Acknowledgements

None.

References

1. Virchow VR. Zur pathologischen anatomic der netzaut und des scherven. *Virchow's Arch Pathol Anat*. 1856;10:170–193.
2. Straatsma BR, Foos RY, Heckenlively JR, Taylor GN. *Am J Ophthalmol*. 1981;91(1):25–38.
3. Tarabisshy AB, Alexandrou TJ, Traboulsi EL. Syndrome of myelinated retinal nerve fibers, myopia and amblyopia: a review. *Surv Ophthalmol*. 2007;52(6):588–596.
4. Fitzgibbon T, Nestorovsky Z. Morphological consequences of myelinisation in the human retina. *Exp Eye Res*. 1997;65:809–819.
5. Straatsma BR, Heckenlively JR, Foos RY, Shahinian JK. Myelinated retinal Fibers associated with ipsilateral myopia, amblyopia and strabismus. *Am J Ophthlmol*. 1979;88:506–510.
6. Gharai S, Prakash G, Ashok Kumar D, Jacob S, Agarwal A, Arora V. Spectral domain optical coherence tomographic characteristics of unilateral peripapillary myelinated retinal nerve fibers involving the macula. *J AAPOS*. 2010;14(5):432–434.
7. Naghib J. Triad of myelinated retinal nerve fibers, axial myopia and amblyopia. *J Ophthalmic Vis Res*. 2010;5(4):284–285.
8. Lam DS, Leung KS, Mohamed S, et al. Regional variations in the relationship between macular thickness measurements and myopia. *Invest Ophthalmol Vis Sci*. 2007;48:376–382.
9. Ellis GS, Frey I, Gouterman RZ. Myelinated nerve fibers, axial myopia and refractory amblyopia: an organic disease. *J Pediatr Ophthalmol Strabismus*. 1987;24:111–119.
10. Kee C, Hwang JM. Visual prognosis of amblyopia associated with myelinated retinal nerve fibers. *Am J Ophthalmol*. 2005;139:259–265.
11. Bodack MI. Medullated Nerve Fibers confounding or contributing to amblyopia – a case report and review. *J Behav Optometry*. 2001;12(5):127–131.
12. Laghmari M, Boutimzine N, Karim A, Daoudi R, Mohcine Z. Fibres à myeline péripapillaires étendues, myopie forte ipsilatérale et amblyopie réfractaire. *J Fr Opthtalmol*. 2004;27(2):188–190.
13. Kreidl KO, Lin DY, Egbert JE. Myelinisation of the macula associated with disabling photophobia. *Arch Ophthalmol*. 2003;121:1204–1205.
14. Rodrigues MI, Loureiro C, Almeida L, Monteiro-Grillo M. Persistência de fibras de mielina e não só: Síndrome de straatsma. *Oftalmologia*. 2013;37(2):117–124.