

# Straatsma syndrome: unilateral myelinated retinal nerve fibre layer, high myopia, strabismus and amblyopia

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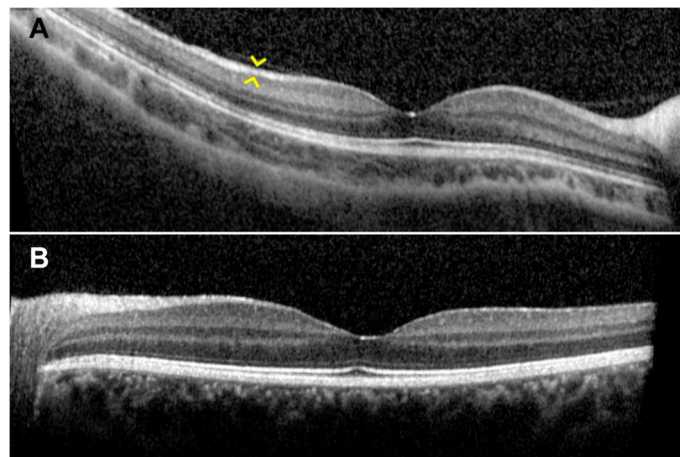
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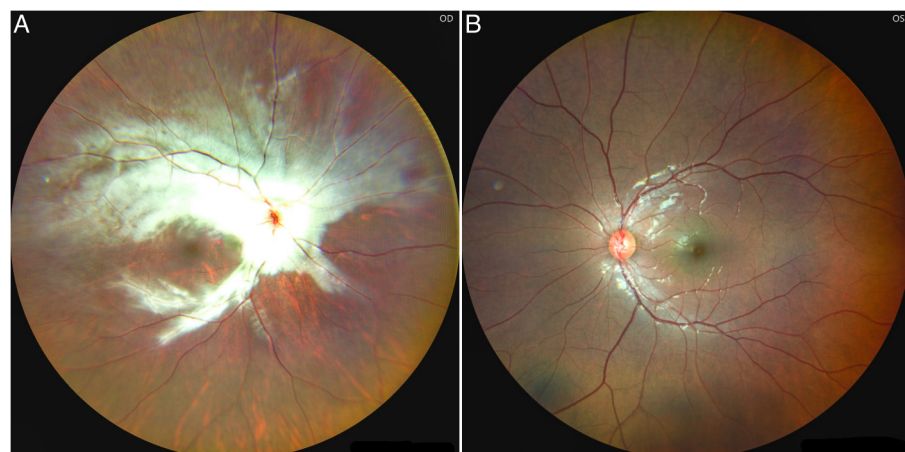
**Figure 2** (A) OCT of the right eye through the macula demonstrating hyper-reflective (arrowheads) retinal nerve fibre layer in the parafoveal area. (B) Left eye OCT showing normal retinal layers. OCT, optical coherence tomography; OD, right eye; OS, left eye.

## 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  $180^\circ$  in the right eye. The patient had right exotropia of 10 prism diopters on modified Krimsky's test with full extraocular motility. He had no stereopsis and demonstrated

## Patient's perspective

I was deeply saddened by the fact that we came to know about my child's condition late enough to be treated effectively. Wearing contact lenses also did not help him much. Had I known about this disease earlier, my child would stand a better chance of regaining his eyesight. I hope there is a way to know about this earlier.



**Figure 1** (A) Fundus photo of the right eye showing extensive myelination of the retinal nerve fibre layer. (B) Normal fundus of the left eye.



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## Learning points

- ▶ Myelination of retinal nerve fibre layer is a benign condition and does not affect vision, but may be associated with high myopia and strabismus.
- ▶ Early recognition of this condition in children by paediatricians and ophthalmologists can prevent dense amblyopia later in life.
- ▶ Treatment consists of full optical correction and initiation of antiamblyopia therapy.

right eye suppression on sensory examination. Dilated fundus examination was performed, which showed the presence of a myelinated retinal nerve fibre 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 27.60 mm and that of the left eye was 22.07 mm. Optical coherence tomography was performed, which depicted parafoveal hyper-reflective retinal nerve fibre 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. There was no improvement in the visual acuity after 6 months of amblyopia treatment and the patient was explained about the possible need for strabismus surgery in the future. Straatsma syndrome is defined as myelinated nerve fibre with myopia and amblyopia.<sup>1</sup> 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 fibre layer.<sup>2</sup> Amblyopia, if detected early in life, can be treated with good visual outcomes.<sup>3 4</sup>

**Contributors** MJ diagnosed the condition, investigated and advised treatment. JMS was involved in collection of images with valid consent. MJ and JMS were involved in drafting the manuscript of the case report. MJ, RA and HKW revised the manuscript and maintained the standards of the manuscript as per BMJ protocols. The final manuscript was approved by all the four authors. All authors are responsible for the integrity and accuracy of the data provided.

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