

Answer to Ophthalmproblem continued from page 355

### 3. Myelinated retinal nerve fibre layer

Myelinated retinal nerve fibre layer (MRNFL) is a retinal lesion caused by the abnormal myelination of the nerve fibres of the retina. The lesion typically appears as striated gray or white opacification with feathery edges, and often follows the distribution of the nerve fibres.<sup>1</sup> The estimated incidence of MRNFL is 0.98%.<sup>1</sup> This paper is an update of a previous paper on MRNFL published by Baxter and Sharma in 2001.<sup>2</sup>

Although the pathogenesis of MRNFL has not been definitively defined, oligodendrocytes have been implicated. Oligodendrocytes are the main supporting cells of the central nervous system and are responsible for insulating long axons with myelin in order to increase the speed of conduction of action potentials. During normal development, the lamina cribrosa—a perforated component of the sclera allowing exit of the retinal nerve fibres from the eye giving rise to the optic nerve—prevents migrating oligodendrocyte progenitor cells from entering the developing eye. This barrier function is thought to be carried out by dense astrocytic processes that aggregate at the lamina cribrosa.<sup>3</sup> Thus, myelination of the optic nerve ends at the lamina cribrosa and the retinal nerve fibres remain unmyelinated. However, in MRNFL, the nerve fibres on the retina are also sheathed in myelin. Histologic examination of MRNFL specimens by Straatsma and colleagues revealed the presence of oligodendrocyte-like cells in the retina.<sup>1</sup> Interestingly, the same study reported normal-looking lamina cribrosa. It is therefore likely that oligodendrocyte progenitors migrate to the retina before the development of the barrier function of the lamina cribrosa or else during a transient period of interruption of this function.<sup>4</sup> Retinal nerve fibre layer myelination can also result from myelinated microglial cells that were activated to the retina in utero.<sup>4</sup> In addition, Parulekar and Elston reported a case of MRNFL caused by an optic nerve glioma that might have compromised the lamina cribrosa.<sup>5</sup>

The effect of MRNFL on visual function is highly variable and dependent on the location of the lesion and its size.<sup>4</sup> Many cases of MRNFL are incidental findings in asymptomatic patients. In contrast, large lesions or those that cover the macula are more likely to produce visual deficits. Moreover, MRNFL can cause axial myopia and amblyopia in children.<sup>4,6</sup> The presence of axial myopia heightens the degree of visual impairment.<sup>4</sup> Myelinated retinal nerve fibre layer can occasionally present with leukocoria, in which case the differential diagnosis must include retinoblastoma.<sup>1,4</sup>

Although MRNFL can be an isolated finding, it can also be associated with ocular and systemic abnormalities. Ocular associations include branch artery or vein occlusions, vitreous hemorrhage, optic nerve hypoplasia, and neovascularization.<sup>4,7-9</sup> Some systemic disorders

linked to MRNFL include neurofibromatosis type 1,<sup>5</sup> craniofacial abnormalities, vitreoretinopathy with skeletal malformations,<sup>4</sup> and basal cell nevus syndrome.<sup>10</sup>

### Management

Asymptomatic MRNFL does not require treatment.<sup>4</sup> For symptomatic MRNFL, treatment should be directed toward any existing associated conditions. For instance, amblyopia should be treated with occlusion therapy,<sup>4,11</sup> although the results are variable and depend on several factors. Best results are obtained when the size of the retinal lesion is small, the difference in refractive power between the 2 eyes (ie, anisometropia) is low, and the macula appears uninvolved upon initial examination.<sup>12</sup> In contrast, the presence of optic nerve dysplasia or strabismus predicts a poor outcome.<sup>4</sup> Axial myopia, if present, should be treated with corrective lenses.<sup>4</sup> Retinovascular events, such as neovascularization or vitreous hemorrhage, should be treated with argon laser photocoagulation to prevent recurrences.<sup>13</sup>

### Conclusion

Myelinated retinal nerve fibre layer is a condition that produces opaque lesions on the retina. It can be asymptomatic, as in the case of our patient, or cause varying degrees of visual impairment. Although there is no treatment for the condition itself, any associated negative prognostic conditions such as axial myopia, amblyopia, and retinovascular lesions should be treated promptly. Asymptomatic patients need only be followed on a routine basis.

**Ms Eneh** is a third-year medical student at Queen's University in Kingston, Ont. **Dr Schweitzer** is a second-year resident in the Department of Ophthalmology at Queen's University. **Dr Sharma** is a Professor and Research Director in the Department of Ophthalmology and a Professor in the Department of Community Health and Epidemiology at Queen's University.

### Competing interests

None declared

### References

1. Straatsma BR, Foos RY, Heckenlively JR, Taylor GN. Myelinated retinal nerve fibers. *Am J Ophthalmol* 1981;91(1):25-38.
2. Baxter S, Sharma S. Myelinated retinal nerve fibre layer [Ophthalmproblem]. *Can Fam Physician* 2001;47:1730,1731.
3. Hunter SF, Leavitt JA, Rodriguez M. Direct observation of myelination in vivo in the mature human central nervous system. A model for the behaviour of oligodendrocyte progenitors and their progeny. *Brain* 1997;120(Pt 11):2071-82.
4. Tarabishy AB, Alexandrou TJ, Traboulsi EI. Syndrome of myelinated retinal nerve fibers, myopia, and amblyopia: a review. *Surv Ophthalmol* 2007;52(6):588-96.
5. Parulekar MV, Elston JS. Acquired retinal myelination in neurofibromatosis 1. *Arch Ophthalmol* 2002;120(5):659-61.
6. Schmidt D, Meyer JH, Brandt-Dohrn J. Wide-spread myelinated nerve fibres of the optic disc: do they influence the development of myopia? *Int Ophthalmol* 1996-1997;20(5):263-8.
7. Leys AM, Leys MJ, Hooymans JM, Craandijk A, Malenfant M, van Germeersch D, et al. Myelinated nerve fibers and retinal vascular abnormalities. *Retina* 1996;16(2):89-96.
8. Mehta JS, Raman J, Gupta N, Sinha A. Retinal vascular anomalies in acquired myelinated nerve fibers. *Acta Ophthalmol Scand* 2003;81(3):311-2.
9. Minning CA Jr, Davidorf FH. Neovascularization associated with myelinated nerve fibers: a case report. *Ann Ophthalmol* 1983;15(2):1142-4.
10. De Jong PT, Bistervels B, Cosgrove J, de Grip G, Leys A, Goffin M. Medullated nerve fibers: a sign of multiple basal cell nevi (Gorlin's) syndrome. *Arch Ophthalmol* 1985;103(12):1833-6.
11. Summers CG, Romig L, Lavoie J. Unexpected good results after therapy for anisometropic amblyopia associated with unilateral peripapillary myelinated nerve fibers. *J Pediatr Ophthalmol Strabismus* 1991;28(3):134-6.
12. Kee C, Hwang JM. Visual prognosis of amblyopia associated with myelinated retinal nerve fibers. *Am J Ophthalmol* 2005;139(2):259-65.
13. Silvestri G, Sehmi K, Hamilton P. Retinal vascular abnormalities. A rare complication of myelinated nerve fibers? *Retina* 1996;16(3):214-8.