CASE REPORT

Syndrome of myelinated retinal nerve fibres, myopia, amblyopia and strabismus in a Nigerian

Vivian B. Osaguona, Odarosa M. Uhumwangho

Department of Ophthalmology, University of Benin Teaching Hospital, Benin City, Edo State, Nigeria

Address for correspondence:
Dr. Vivian B. Osaguona, Department of Ophthalmology, University of Benin Teaching Hospital, P.M.B. 1111, Benin City, Edo State, Nigeria.
E-mail: vivian_osaguona@yahoo.co.uk

ABSTRACT

Myelinated retinal nerve fibres (MRNF) are rare congenital anomalies. They may present in a syndrome characterised by ipsilateral myelinated retinal nerve fibres, myopia and amblyopia. We report a case of this rare condition with unilateral extensive MRNF, axial myopia, amblyopia and strabismus in a Nigerian girl.

Key words: Amblyopia, axial myopia, myelinated retinal nerve fibres, Nigeria, strabismus

INTRODUCTION

Myelinated retinal nerve fibres (MRNF) are rare congenital anomalies that appear as grey-white opaque lesions on the retina with feathery edges that obscure retina details. They may be located on the disc or elsewhere on the retina. However, few reports of acquired cases have been documented.[1] Straatsma *et al.*,[2] in a study of 3968 autopsies found MRNF in 39 (0.98%) cases; four of the patients had a syndrome consisting of ipsilateral myopia, amblyopia, strabismus and MRNF.

Majority of MRNF are unilateral[2-4] and usually remain stable overtime, but there are reported cases of progression and even disappearance of MRNF following other ocular pathologic processes and treatment procedures.[4]

CASE REPORT

A 22-year-old Nigerian female student was referred to us on account of poor vision in the left eye, first noticed incidentally 12 years ago, on closing the right eye. She had no other ocular symptoms, but had a history of absence seizures.

Unaided visual acuity was 6/5 right eye and counting fingers at 1metre in the left eye. The right eye was essentially normal. The left eye had an exotropia of 15° with full ocular motility, normal anterior segment and a subtle relative afferent pupillary defect. Intraocular pressure was 10mmHg by applanation tonometry in either eye. On dilated fundoscopy, the right eye had a normal disc with a cup to disc ratio of 0.3, and normal retina. In the left eye, there was extensive MRNF extending from the disc along the superotemporal arcade up to the midperiphery, sparing the macula [Figure 1].The axial lengths were 23.6mm and 28.7mmm in the right and left eye respectively. Refraction revealed emmetropia in the right eye and −18.75 −1.75 × 153° in the left eye. There was no improvement in vision with optical correction in the left eye suggesting amblyopia.

DISCUSSION

In the normal eye, the retinal nerve fibre layer is transparent and unmyelinated, allowing visualisation of the retinal blood vessels. Myelinated retinal nerve fibres are diagnosable clinically. They are recognised

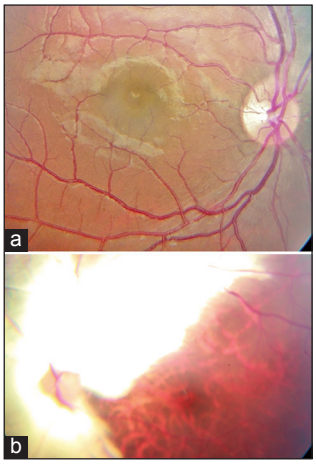

Figure 1: (a) normal fundus in the right eye (b) extensive myelinated retinal nerve fibre layer in the left eye

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Quick Response Code:

Website: www.nigeriamedj.com

DOI: 10.4103/0300-1652.144714

on fundoscopy as grey-white opaque patches on the retina with striations and feathery edges, which usually give away the diagnosis. They obscure the retinal blood vessels and the disc. In majority of cases, the patient's vision is not affected; however, some cases have been associated with the refractive error, myopia, which causes poor vision.[4] The patient may have myopia in the eye with MNRF, and emmetropia (no refractive error) or myopia of a lesser degree in the fellow eye. This is referred to as anisometropic myopia. The myopia may be unresponsive to optical correction with lenses (that is the vision cannot be improved with optical correction), in which case the patient is referred to as having amblyopia. There may also be associated misalignment of the visual axes of the eyes referred to as strabismus (squint), in which there may be an outward deviation of the eye (exotropia) or an inward deviation (esotropia).[3-6] The patient, therefore, has a syndrome of ipsilateral myelinated retinal nerve fibres, anisometropic myopia , amblyopia and strabismus.

Although, MRNF is known to be associated with myopia, rare cases of MRNF associated with hypermetropia and amblyopia have been documented.[7,8] Other ocular and systemic associations with MRNF include epiretinal membrane, branch retinal artery occlusion, branch retinal vein occlusion, neovascularisation, recurrent vitreous haemorrhage, keratoconus, neurofibromatosis 1 and Gorlin's syndrome.[4,9]

The exact pathogenesis of MRNF is not known. Oligodendrocytes are responsible for the myelination of the ganglion cell axons, which normally begins at the lateral geniculate body and proceeds anteriorly to end at the lamina cribosa, which is thought to act as a barrier to the anterior migration of myelination into the retinal nerve fibres.[4,10] It has been postulated that MRNF results from an imbalance between the formation of the lamina cribosa, which proceeds posteriorly from the limbus and the process of myelination, which begins from the lateral geniculate body.[10] This imbalance is believed to be more likely to occur in myopia, due to the enlarged eyeball.[10] In addition, a temporary loss in the barrier function of the lamina cribosa may result in abnormal dislocation of oligodendrocyte–like cells into the retinal nerve fibres.[4]

Our patient had a 15° exotropia and a subtle relative afferent pupillary defect. Strabismus, when present in MRNF with anisometropic amblyopia, could either be an exotropia or an esotropia,[3-6] however, a relative afferent pupillary defect is not a common finding.[4]

The visual dysfunction in this syndrome may be attributable either to amblyopia or to an organic cause from retinal or optic nerve abnormalities.[4,10] Therapeutic options for this condition include corrective lenses for myopia and aggressive amblyopic therapy.[4] However, variable visual outcomes with amblyopic therapy have been reported with a few patients having an improved visual outcome up to 20/30 or better, while most cases remained refractory to treatment.[3,4,10] Larger areas of retinal nerve fibre myelination and higher degrees of anisometropia are associated with poorer visual prognosis with amblyopic therapy.[3]

In conclusion, myelinated retina nerve fibres are rare and may be associated with ipsilateral myopia, amblyopia and strabismus. The exact pathogenesis remains unknown and the visual prognosis is guarded.

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How to cite this article: Osaguona VB, Uhumwangho OM. Syndrome of myelinated retinal nerve fibres, myopia, amblyopia and strabismus in a Nigerian. Niger Med J 2014;55:517-8.

Source of Support: Nil, **Conflict of Interest:** None declared.