

Bilateral Myelinated Retinal Nerve Fibres Associated With Ocular Hypertension and Exotropia: A Rare Clinical Entity

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Abstract: Myelinated retinal nerve fibres are rare congenital, non progressive anomalies that appear as grey white opaque lesions with feathery edges that obscure retinal details. These lesions are asymptomatic and are detected incidentally on routine ophthalmological examination. Although generally they are considered to be benign fundoscopic finding, myelinated nerves have been associated with visual field defects, severe myopia, amblyopia, anisometropia and strabismus. We report a 55 year old male patient with bilateral, multiple distinct myelinated nerve fibres seen in association with ocular hypertension and comitant exotropia.

Keywords: Alternating exotropia, Ocular hypertension, Myelinated retinal nerve fibres.

I. Introduction

Myelinated retinal nerve fibre layers also known as medullated retinal nerve fibres are white, well demarcated fan shaped patches in the distribution of the RNFL. Myelinated retinal nerve fibres were first described by Virchow in 1856¹. In a retrospective study on the prevalence and the location of myelinated retinal nerve fibres by Kodama and associates², the prevalence of myelination of the RNFL was found to be 0.57%. Myelinated nerve fibres can be bilateral in 7.7% of cases³. In current theory of pathogenesis, myelinated retinal nerve fibres is thought to be due to presence of ectopic oligodendrocyte-like cells in the retina as a result of a developmental or acquired insult⁴. Most of the cases of myelinated retinal nerve fibres are sporadic. However familial cases of myelinated retinal nerve fibres have been reported both in isolation and in combination with the systemic syndromes like:

1. Growth retardation, alopecia, pseudoanodontia, optic atrophy (GAPO syndrome) with end stage glaucoma⁵.
2. Vitreoretinopathy and skeletal malformations⁶.
3. Turner syndrome⁷.
4. Down's syndrome⁸.

Ophthalmoscopically, myelinated retinal nerve fibres appear as white or grey white fan shaped striated patches in the distributions of the RNFL, which obscure the underlying retinal vessels. They may be single or multiple and can vary in size from small (approx 1 Disc Diameter) to large lesions. In about 33% of cases myelinated retinal nerve fibre patches are contiguous with the optic nerve, while in 66% cases myelinated retinal nerve fibres can be seen as patches discontinuous from the optic nerve head. In about 12% of the cases, myelinated retinal nerve fibres can appear as multiple distinct lesions.³

II. Case History

A 55 year old male patient presented to our clinic with complaints of defective vision for distance and near of 6 months duration. His ocular examination revealed a Un Corrected Visual Acuity of 6/18 in both eyes corrected to 6/6 with -1.0DS with +2.5DS addition for near vision. The patient had 30° alternating exotropia (Fig-1) and his anterior segment examination was normal. His presenting intraocular pressure (IOP) was 26 mm Hg (Right Eye) and 24 mm Hg (Left Eye) by Goldmann Applanation Tonometry. A diurnal IOP monitoring was done which showed a maximum IOP of 26 mm Hg Right Eye and 27 mm Hg Left Eye and a minimum IOP of 18 mm Hg (Right Eye) and 19 mm Hg (Left Eye). His posterior segment evaluation showed a normal optic disc with multiple patches of myelinated nerve fibres distributed in the superior and inferior nasal quadrants of varying sizes with normal temporal retina (Fig-2 Right Eye, Fig-3 Left Eye). His visual field examination (central 30-2) was within normal limits.

Based on the above findings, a diagnosis of ocular hypertension was made and the patient was started on Timolol maleate 0.5% eye drops to be used twice daily. His IOP readings taken one month after starting the treatment has been 18 mm Hg (Both Eyes) by Goldmann Applanation Tonometry.

III. Discussion

Myelinated retinal nerve fibres have been referred to as a 'papillae leporina' and are incidentally detected as asymptomatic white grey lesions obscuring the retinal details. When making the initial diagnosis it is very important to differentiate myelinated retinal nerve fibres which is typically a benign condition from other potentially serious conditions like:

1. Cotton wool spots (seen in Diabetic Retinopathy, Hypertensive Retinopathy, Papilloedema)
2. Retinal infiltrates which could be infectious, inflammatory or neoplastic.
3. Retinoblastoma
4. Branch Retinal Artery Occlusion.

The most common ocular associations of myelinated nerve fibres are strabismus (exotropia and esotropia) found in 66% of patients and amblyopia and anisometropia⁶. Myelinated nerve fibres are most often congenital and non progressive, but few cases of acquired progressive lesions in childhood and adulthood have been reported. Causes of acquired and progressive myelination of the RNFL:

1. Blunt trauma⁹
2. Optic nerve sheath fenestration for chronic papilloedema¹⁰
3. Arnold-chiari malformation associated with hydrocephalus¹¹
4. Von Recklinghausen's disease¹²

Myelinated RNFL have been reported to disappear in association with conditions like

1. Primary Open Angle Glaucoma¹³
2. Branch Retinal Artery Occlusion¹⁴
3. Optic neuritis¹⁵
4. Diabetic retinopathy¹⁶

Figures



Fig-1 Photograph of the patient showing alternative exotropia





Fig-2. Right Eye fundus photographs showing a normal optic disc with multiple patches of myelinated nerve fibres distributed in the superior and inferior nasal quadrants of varying sizes with normal temporal retina

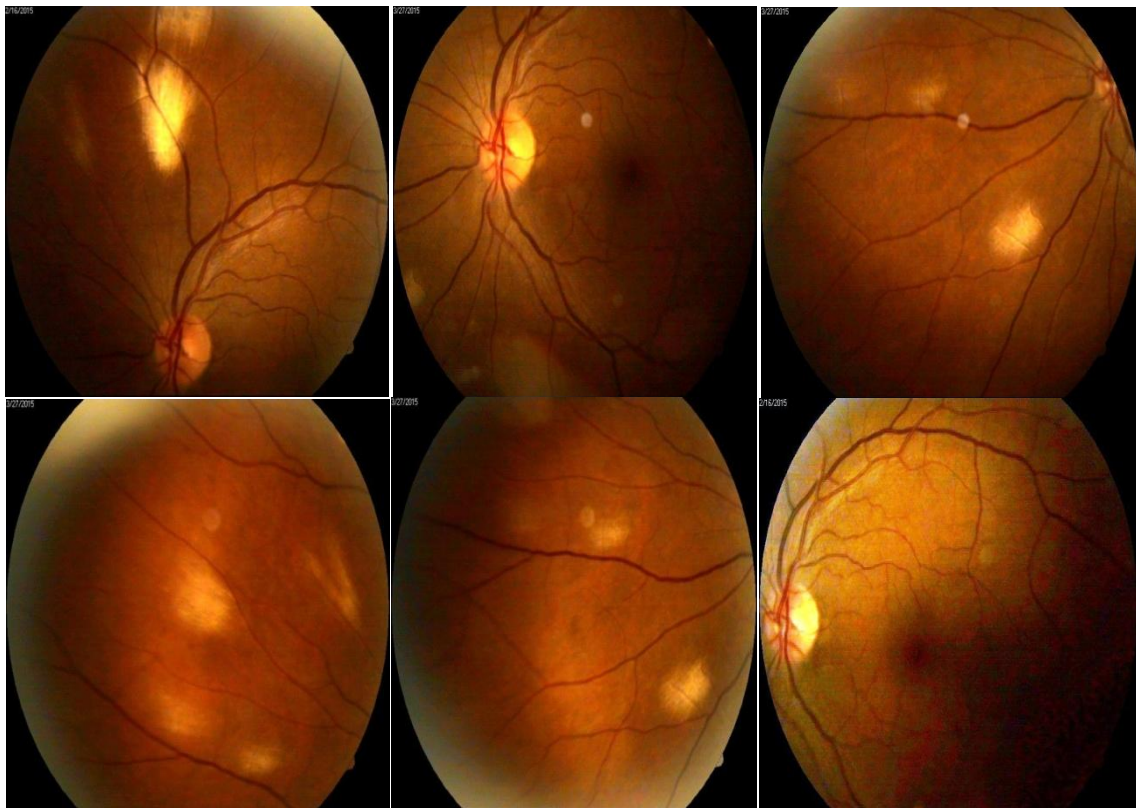


Fig-3. Left Eyefundus photographs showing a normal optic disc with multiple patches of myelinated nerve fibres distributed in the superior and inferior nasal quadrants of varying sizes with normal temporal retina

IV. Conclusion

Myelinated retinal nerve fibres are benign and innocuous lesions that are detected incidentally on ocular examination, The various ocular associations and differential diagnosis have to be kept in mind in patients who are detected to have myelinated retinal nerve fibres

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