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Bilateral myelinated retinal nerve fibers: A rare benign anomaly

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Abstract
Myelinated retinal nerve fibers (MRNF) represent a rare benign developmental anomaly resulting from continuation of myelination beyond the lamina cribrosa. While typically unilateral, bilateral presentation is extremely uncommon. We report a case of a 27-year-old male who presented with diminution of vision for distant objects in both eyes for three years. Comprehensive ophthalmic examination revealed best-corrected visual acuity of 6/6 in both eyes with characteristic fundoscopic findings of bilateral myelinated nerve fibers appearing as white peripapillary patches obscuring the optic disc margins. Optical coherence tomography confirmed the diagnosis by demonstrating hyperreflective thickening of the retinal nerve fiber layer. This case emphasizes the importance of recognizing this benign condition to avoid unnecessary investigations and interventions while ensuring appropriate follow-up for associated refractive errors and visual field defects.

Keywords: Myelinated retinal nerve fibers, bilateral, optic disc anomaly, visual field defect, myopia

Introduction
Myelinated nerve fibers of the retina represent a benign developmental anomaly first described by Virchow in 1856 [1]. During normal prenatal development, myelination of the optic nerve commences at the lateral geniculate body and progresses anteriorly, typically terminating at the lamina cribrosa around birth. Myelinated retinal nerve fibers (MRNF) occur when this physiological barrier fails, allowing oligodendrocyte migration into the retina with subsequent myelin sheath formation around retinal ganglion cell axons [2]. The condition is typically unilateral with an estimated prevalence of 0.3-0.6% in the general population [3]. Bilateral involvement is considerably rarer, occurring in approximately 6-7% of all MRNF cases [4]. We present a rare case of bilateral MRNF to highlight its clinical features and diagnostic approach.

Case Report
A 27-year-old male, Abdul Rauf, presented with diminution of vision for distant objects in both eyes for three years. Visual impairment was gradual in onset and progressive, without pain, redness, or discharge. There was no history of ocular trauma, surgery, or spectacle use. Systemic history was non-contributory.

Clinical Examination
Uncorrected visual acuity was 6/24 in the right eye and 6/9 in the left eye, improving to 6/6 bilaterally with refractive correction (compound myopic astigmatism). Anterior segment examination was unremarkable bilaterally. Intraocular pressure by iCare tonometry measured 15 mmHg right eye and 16 mmHg left eye. Color vision using Ishihara plates was 20/22 bilaterally. Amsler grid testing was normal. Confrontation visual field testing revealed bilateral inferior field defects.

Fundus Examination
Dilated fundus examination revealed extensive peripapillary white patches with feathery borders obscuring the optic disc margins bilaterally. The characteristic white, striated appearance following the arcuate nerve fiber layer pattern was consistent with myelinated nerve fibers. Retinal vessels appeared normal. The macula showed normal foveal reflex, and peripheral retina was unremarkable. Media was clear in both eyes.

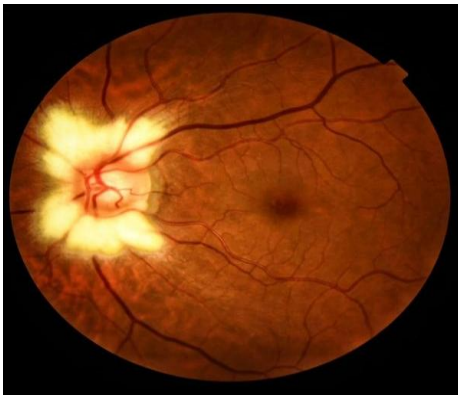

Fig 1: Fundus photograph of the left eye showing extensive peripapillary myelinated nerve fibers appearing as white striated patches obscuring the optic disc margin with feathery borders following the nerve fiber layer distribution

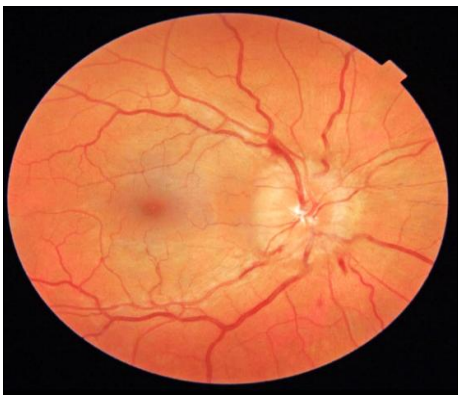

Fig 2: Fundus photograph of the right eye demonstrating similar peripapillary myelinated nerve fibers with characteristic white appearance and irregular margins extending into the retinal nerve fiber layer

Investigations

Optical coherence tomography (OCT) revealed hyperreflective thickening of the retinal nerve fiber layer corresponding to the white patches, consistent with myelin presence. Underlying retinal architecture was preserved with normal ganglion cell complex thickness in uninvolved areas.

Diagnosis and Management

Based on the characteristic clinical appearance, bilateral presentation, and OCT findings, bilateral myelinated retinal nerve fibers were diagnosed. With best-corrected visual acuity of 6/6 bilaterally and no complications, no active intervention was required. Refractive correction was prescribed. The patient was counseled regarding the benign nature of the condition and advised annual follow-up to monitor optic nerve status, visual field stability, and refractive error progression.

Discussion

While the exact etiology of MRNF remains uncertain, theories include genetic predisposition, failure of the lamina cribrosa as a barrier, and premature oligodendrocyte migration [2, 3]. Some reports suggest familial clustering, while others document acquired cases following inflammatory or traumatic events [5]. The bilateral presentation in our case is particularly noteworthy given its rarity. Straatsma *et al*. reported only 6.25% of MRNF cases were bilateral [4], suggesting possible underlying genetic or developmental mechanisms affecting both eyes.

The clinical appearance of MRNF is distinctive, typically allowing diagnosis on fundoscopy alone. The white or gray-white patches with feathery, striated borders respecting the nerve fiber layer architecture are pathognomonic. Differential diagnoses include cotton-wool spots, retinal astrocytic hamartomas, hard exudates, and infiltrative processes [6]. Cotton-wool spots lack the striated appearance and resolve over weeks to months, whereas MRNF remain stable. OCT confirms diagnosis by demonstrating hyperreflective nerve fiber layer thickening without retinal edema [7].

MRNF association with refractive errors, particularly myopia, is well documented. Tarabishy *et al*. described a syndrome comprising myelinated retinal nerve fibers, myopia, and amblyopia [1]. Our patient demonstrated compound myopic astigmatism consistent with this association. Visual field defects corresponding to myelination areas are common, resulting from axonal dysfunction or adjacent nerve fiber layer thinning [8]. Our patient's inferior field defects correlated with superior peripapillary myelinated fiber distribution.

Most MRNF cases remain stable throughout life. However, rare complications include vitreous hemorrhage, retinal detachment, and progressive visual loss [5, 9]. Regular follow-up is warranted to detect complications early. Patient education forms the cornerstone of care. Refractive correction should be optimized, particularly in pediatric cases for amblyopia prevention. There is no role for surgical or medical intervention in uncomplicated cases.

Conclusion

Bilateral myelinated retinal nerve fibers represent a rare benign anomaly that may present with refractive errors and visual field defects despite excellent visual acuity. The characteristic fundoscopic appearance allows clinical diagnosis confirmed by OCT. Recognition of this condition prevents misdiagnosis and inappropriate management. Regular follow-up is recommended to monitor for rare complications. This case emphasizes the importance of comprehensive examination in evaluating optic disc anomalies.

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