Case Report

Progressive Loss of Myelinated Retinal Nerve Fibers in a Case of Open-Angle Glaucoma

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Abstract

Myelinated retinal nerve fibers (MRNFs) result from a developmental anomaly in which ectopic oligodendrocytes myelinate retinal ganglion cell fibers. These fibers typically remain stable over time in the absence of pathology. We identified unilateral MRNFs in a patient during an ophthalmological examination associated with ocular hypertension. Over an 8-year follow-up period, we observed a progressive decrease in the retinal nerve fiber layer and a rarefaction of MRNFs, which we attributed to the lack of regular follow-up and treatment compliance. The absence of neuropathy control in this patient makes this description close to the natural evolution of the disease. Similar progressive disappearance of MRNFs has been observed in cases of Behçet's disease, pituitary adenoma, and open-angle glaucoma. In patients with known MRNFs, their disappearance should alert clinicians to potential optic nerve damage and prompt further examinations to determine the underlying cause.

Keywords: Open-angle glaucoma, myelinated retinal nerve fibers, glaucomatous optic neuropathy

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Introduction

Myelinated retinal nerve fibers (MRNFs) appear as white striated patches with poorly defined borders. This rare and predominantly benign congenital anomaly results from ectopic oligodendrocytes myelinating retinal ganglion cell fibers. Their prevalence is low, ranging from 0.34% to 1.03%,[1,2] and they tend to remain stable over time in the absence of associated pathology.[3] In the scientific literature, MRNFs are most frequently associated with strabismus, anisometropia, and/or amblyopia.[4,5,6]

MRNFs do not impact vision but can interfere with optical coherence tomography (OCT) acquisition due to the masking effect of myelin. When extensive, the software can misinterpret them as a large papilla, resulting in incorrect automatic centering.[7] Their presence can delay the diagnosis of open-angle glaucoma, as reported in a clinical case where the entire optic nerve was covered.[7] Although MRNFs are not associated with an increased risk of glaucomatous neuropathy, a syndrome has been reported in children of consanguineous parents.[8] This report presents a unique case of progressive glaucomatous neuropathy associated with MRNF rarefaction and explores its clinical implications.

Case Report

A 47-year-old woman presented for a renewal of optical correction. Her ophthalmological history included strabismus surgery on the left eye during childhood. Examination revealed a best corrected visual acuity of 20/20 in both eyes. Intraocular pressure (IOP) measured by air puff tonometry was normal in the right eye (13 mmHg; pachymetry of 516 µm) but showed moderate ocular hypertension in the left eye, confirmed by Goldmann applanation tonometry (23 mmHg; pachymetry of 518 µm). Fundus examination revealed MRNFs in the inferior optic nerve region of the left eye. Anterior segment examination was normal in both eyes, and the iridocorneal angle was open.

OCT (Cirrus HD-OCT 4000; Carl Zeiss Meditec, Dublin, CA, USA) showed a normal retinal nerve fiber layer (RNFL) but a slight decrease in ganglion cell layer thickness in the left eye (Figure 1). Three months later, unilateral ocular hypertension was reconfirmed by Goldmann applanation tonometry (10 mmHg in the right eye and 21 mmHg in the left eye). Visual field testing (Octopus Interzeag 1-2-3, Interzeag, Schlieren, Switzerland) was inconclusive, even after repeated attempts. Consequently, a latanoprost eye drop (Monoprost 50 µg/mL; Théa Pharma, Clermont-Ferrand, France) was prescribed at the end of the consultation. A month later, another visual field test revealed a minor nasal defect in the left eye, but the patient did not return for follow-up interpretation with the ophthalmologist.

Eight months later, the patient presented with conjunctivitis at another ophthalmology center, without mention of her recent examinations. IOP without medication, measured by air puff tonometry, was normal in the right eye (14 mmHg) but elevated in the left eye (26 mmHg). OCT (Topcon 3D OCT-1 Maestro, Oakland, NJ, USA) showed normal RNFL thickness in the right eye but a small inferior temporal defect in the left eye. Eleven months later, IOP in the left eye remained elevated (27 mmHg), with thinning of the RNFL in the inferior temporal region. Local treatment was prescribed for the left eye (latanoprost 50 µg/mL; Monoprost). At the check-up appointment, her IOP had decreased to 21 mmHg and the patient was informed of the risks associated with noncompliance with treatment. Investigations were conducted to explore secondary causes of intraocular hypertension in the context of this unilateral affection.

After 1 year, the target IOP (>18 mmHg) was not achieved (22 mmHg in the left eye). Progressive RNFL thinning was observed in both the superior and inferior quadrants. Glaucomatous optic neuropathy was confirmed, with fundus photographs showing rarefaction of MRNFs (Figure 2). Despite a reminder of the importance of regular control and compliance with treatment, the patient's visits remained irregular. Ten months later, the IOP in the left eye was 21 mmHg, due to the self-reported non-compliance with treatment.

The patient was referred to the initial ophthalmology center for consultation with a glaucoma specialist. IOP was measured as 26 mmHg in the left eye (Goldmann applanation tonometry). OCT (Cirrus HD-OCT 5000; Carl Zeiss Meditec, Dublin, CA, USA) was not interpretable due to miosis, and the patient declined an examination with mydriatic eye drops. Due to the failure of previous treatments, combination therapy with

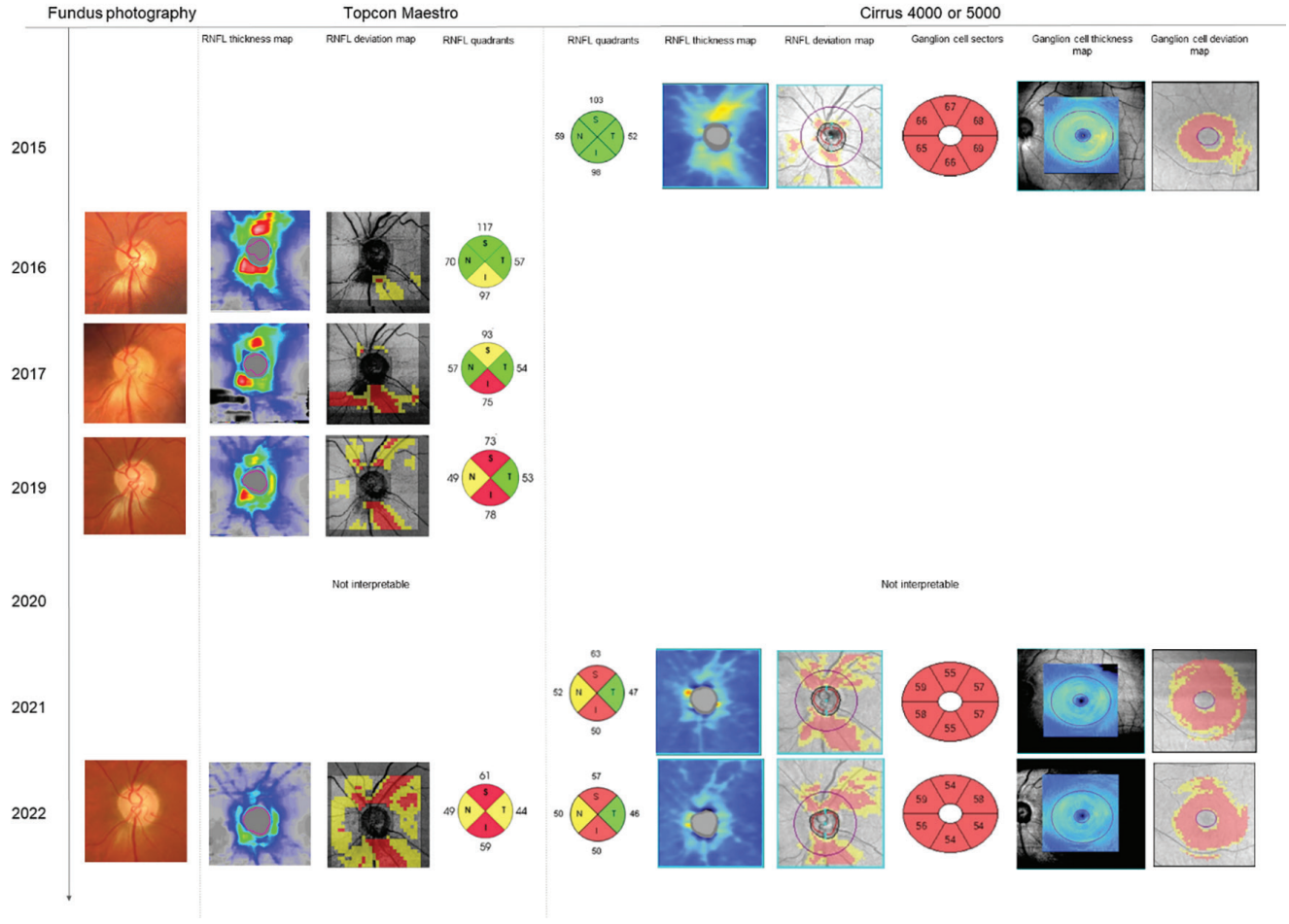

Figure 1. Evolution of the retinal nerve fiber layer and ganglion cells of the patient during follow-up

latanoprost 50 µg/mL (Monoprost) and dorzolamide 20 mg/mL + timolol 5 mg/mL (Dualkopt; Théa Pharma, Clermont-Ferrand, France) was initiated.

Ten months later, target IOP was still not achieved (19 mmHg in the left eye). RNFL thinning progressed in the superior and inferior quadrants. The patient declined selective laser trabeculoplasty and modifications to treatment due to compliance issues. Eight months later, IOP decreased to 17 mmHg in the left eye. Topcon Maestro OCT and fundus photography were performed to compare results with those from the previous clinic.

Discussion

This case highlights the progressive rarefaction of MRNFs in association with uncontrolled glaucomatous optic neuropathy, documented through fundus photography and OCT. The poor treatment compliance of the patient allowed for an observation close to the natural progression of the disease, providing unique insights into MRNFs and glaucoma. The unilateral manifestation of glaucomatous damage enables a comparison with the stability of the RNFL in the fellow eye.

To our knowledge, the regression of MRNFs has been reported in only two other articles.[9,10] In the first case, the patient

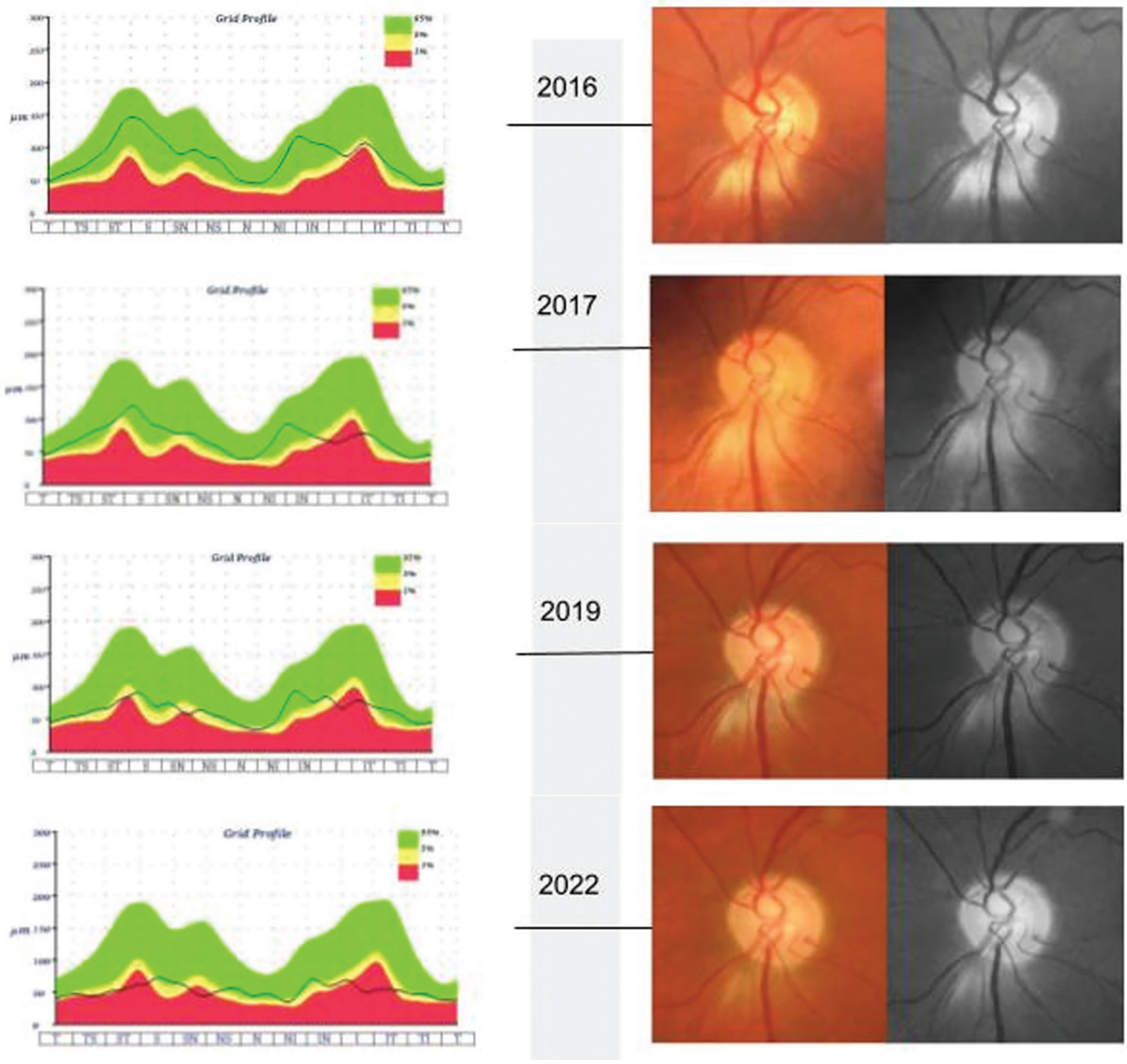

Figure 2. Evolution of the retinal nerve fiber layer and comparison with fundus photography on the Topcon Maestro (3D OCT-1 Maestro, Oakland, NJ, USA)

had a history of diabetes, high myopia, amblyopia, and bilateral glaucoma with high IOP (>37 mmHg).[10] Sellem and Poli[9] also described a patient with unilateral ocular hypertension and MRNFs, similar to this case. However, the current case provides a more detailed description of the progressive RNFL and MRNF rarefaction.

The disappearance of known MRNFs should alert clinicians to possible optic nerve damage. Various potential etiologies exist, including ischemic attacks, Behçet's disease, pituitary adenoma, retinal astrocytoma, Gorlin syndrome, and open-angle glaucoma.[9,10,11,12,13,14,15] These conditions share a common pathway of ganglion cell layer damage. Additional investigations are essential to rule out secondary causes in cases of unilateral ocular hypertension with MRNFs. The association between RNFL thinning and anatomic rarefaction of MRNFs should alert clinicians to potential progressive optic neuropathy.

Ethics

Informed Consent: The patient's consent has been obtained.

Declarations

Authorship Contributions

Surgical and Medical Practices: T.L., P.G., Concept: T.J., C.Z., Design: C.Z., Data Collection or Processing: T.J., Analysis or Interpretation: T.L., P.G., Literature Search: T.J., Writing: T.J., T.L., C.Z.

Conflict of Interest: No conflict of interest was declared by the authors.

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