squamous cell carcinoma both on the bulbar conjunctiva (Figure, E) as well as in the anterior chamber angle and iris (Figure, F). The best-corrected visual acuity remains stable at 20/200 in the treated eye.

Comment. Intraocular invasion of conjunctival squamous cell carcinoma is rare. The Bowman membrane is a thick acellular layer composed largely of collagenous tissue. Due to its structure, it often acts as an impenetrable barrier to the intraocular spread of conjunctival melanoma. More aggressive histologic variants that have a greater potential for invasion include spindle cell and mucocysticoid carcinomas. It is of note that although the lesion in our patient did not fall within this subtype, it still demonstrated invasive behavior. Although squamous cell carcinoma generally exhibits slow growth and has a low likelihood of metastasis, this case highlights the need for accurate diagnosis and early intervention.

This patient received successful treatment of invasive squamous cell carcinoma of the conjunctiva with proton beam therapy. Recently, the therapeutic options available to the clinician for the treatment of superficial conjunctival and corneal squamous cell carcinoma have expanded. Promising reports in the literature describe the use of photodynamic therapy and topical mitomycin C for the treatment of more extensive and recurrent lesions. However, previous to this article, the options that were suggested for the treatment of intraocularly invasive squamous cell carcinoma were limited to enucleation. This article suggests proton beam therapy as a potential alternative to enucleation.

On review of the literature, some authors describe visually limiting complications with the use of external beam radiation for the treatment of conjunctival malignancy. Compared with other forms of irradiation, proton beam therapy can be delivered precisely to the superficial tissues and anterior chamber structures without significant deeper tissue penetration, thus minimizing unnecessary irradiation of the lens, ciliary body, and retina. Such a treatment approach is particularly suited to elderly patients who may be limited in surgical options by other medical comorbidities. Proton beam therapy was extremely well tolerated by our patient, who experienced no adverse effects from the radiation. Adverse effects may include dry eye, intraocular inflammation, conjunctival scarring, or glaucoma.

Our knowledge of the efficacy of proton beam therapy is limited by the fact that we have only treated 1 patient and that we have relatively short-term follow-up data. The regression of tumor and lack of recurrence in this patient does, however, suggest that proton beam therapy should be considered as a possible alternative to enucleation for the treatment of invasive conjunctival squamous cell carcinoma.

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Unilateral Retinal Nerve Fiber Myelination With Contralateral Amblyopia

During normal prenatal development, retinal ganglion axon myelination starts centrally in the lateral geniculate body and proceeds anteriorly to the optic tracts, chiasm, and nerves. This process of myelination normally terminates shortly after birth at the level of the lamina cribrosa; however, occasionally myelination occurs in the retinal nerve fiber layer. Although generally considered to be a benign funduscopic finding, myelinated nerves have been associated with visual field defects, severe myopia, amblyopia, and strabismus.

To our knowledge, all of the reported cases of amblyopia have occurred in the eye affected with myelinated fibers. In this article, we describe 2 patients with unilateral myelinated nerve fibers who had amblyopia in the eye not affected with nerve fiber layer myelination. We believe that these cases shed light on the causes of reduced visual acuity in patients with retinal nerve fiber layer myelination.

Report of Cases. Case 1. A 5-year-old boy was referred for evaluation of a “wandering eye” that was first noted in infancy. He had received no previous eye treatment. Visual acuity without correction was 20/125 OD and 20/300 OS. Cycloplegic refraction revealed −2.50 diopters (D) OD and +7.50 D OS, correcting visual acuity to 20/40 OD and 20/160 OS. A 23-prism diopter, left-sided
exotropia was present. There was no afferent pupillary defect. Funduscopic examination of the right eye showed extensive myelinated nerve fibers continuous with the optic nerve (Figure 1). The macula appeared normal. The left fundus was unremarkable. Axial length measurements were 25.22 mm OD and 21.32 mm OS. Keratometry showed regular astigmatism: right eye, 40.18 × 41.56 D (mean, 40.87 D); left eye, 39.66 × 41.56 D (mean, 40.69 D). Treatment with glasses and patching of the right eye 6 hours per day was recommended. At the last follow-up visit 10 months after the initiation of amblyopia treatment, his best-corrected visual acuity had remained 20/40 OD and improved to 20/50 OS.

Case 2. A 9-year-old boy was examined after failing a school vision screening. He had no previous ocular examinations or treatment. His uncorrected visual acuity was 20/20 OD and 20/300 OS. Cycloplegic retinoscopy showed +1.00 D OD and +2.75 +1.50 × 90° OS. There was no afferent pupillary defect. Funduscopic examination showed peripapillary retinal nerve fiber layer myelination in the right eye; the left eye showed no abnormality (Figure 2). Axial length measurements were 23.02 mm OD and 21.80 mm OS. Keratometry showed regular astigmatism: right eye, 41.93 × 42.99 D (mean, 42.46 D); left eye, 41.51 × 43.72 D (mean, 42.62 D). Eyeglasses were prescribed and occlusion therapy of 4 hours on weekdays and full-time on weekends was started. At the most recent follow-up visit after 8 months of treatment, visual acuity had improved to 20/100 OS.

Comment. On funduscopic examination, myelinated retinal nerve fibers appear as white to gray-white patches corresponding to the distribution of the ganglion fibers. Lesions are often found incidentally or in patients referred for evaluation of visual disturbances or leukocoria. Frequently, visual acuity is not affected. Straatsma et al found myelinated fibers in roughly 1% of 3968 consecutive autopsies. Myelin was continuous with the optic nerve head in 81% and discontinuous in 19%.

The normal retina is not myelinated because it lacks oligodendroglia, the cells responsible for myelination in the central nervous system. Anomalous distribution of oligodendroglia in the retina is believed to be responsible for retinal nerve fiber myelination. Reported cases demonstrating intact lamina cribrosa, considered to be a barrier to myelination, and the observation of myelination discontinuous with the optic nerve head refute speculation that myelination simply continues anteriorly through the posterior sclera.

Figure 1. Fundus photographs of patient 1 showing extensive myelination of the retinal nerve fiber layer of the right eye (A) and no myelination in the left eye (B).

Figure 2. Fundus photographs of patient 2 showing peripapillary myelination of the nerve fiber layer of the right eye (A) and no myelination in the left eye (B).
It is not known whether myelination of retinal fibers leads to myopia or myopia leads to myelination of retinal fibers. Alternatively, myopia and retinal myelination may be associated with other factors. Schmidt et al proposed that myelinated fibers could blur retinal images and induce visual deprivation. Such deprivation at a critical stage of ocular development could lead to axial elongation, akin to structural changes seen in eyes with a unilateral congenital cataract or severe ptosis. However, scotomas in myelinated retinas are generally smaller than predicted by the extent of myelination, suggesting that light penetrates to the photoreceptor layer despite the myelin interference. Furthermore, many cases of myelinated fibers do not show visual deficits or have significant myopia.

It is also possible that the increased axial length of the eye predisposes to retinal myelination. If the process of lamina cribrosa development is prolonged, as could occur in axial myopia, myelination could continue down the optic nerve and into the retina. Although this theory has some merit, it cannot explain lesions discontinuous with the optic nerve head or myelination of eyes without increased axial lengths.

It is likely that our patients were genetically predisposed to be hyperopic. During ocular development, one eye became myelinated as a cause of, or resulting in, disruption of the programmed growth pattern of that eye. The resultant axial elongation led to 10 D of anisometropia in the first case and more than 2 D in the second case. As a consequence, these patients developed amblyopia in their more ametropic eyes. Myelination did not cause organic loss of vision from macular dysfunction as reported in some of the patients of Hittner and Antoszyk. The question as to whether myelination causes myopia or vice versa is left unresolved, but our cases do suggest that anisometropia can be a more powerful influence on the relative visual acuity of each patient’s eyes than the presence of retinal myelin. Lempert has proposed smaller optic disc size as an organic as opposed to functional reason for decreased vision in hyperopic anisometropia. Although the optic discs of our patients appeared normal in each eye, we cannot exclude this as an alternative explanation for their decreased visual acuity.

With an estimated amblyopia prevalence of 2% to 3%, much of which is anisometropic amblyopia, and a 0.9% prevalence of unilateral nerve fiber layer myelination, one would expect that the combination of these findings would be seen more frequently. We do not know why there have been no previous published reports about this finding, but do believe that these cases are unique in the literature on myelinated retinal nerve fibers.

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Corneal Copper Deposition Secondary to a Variant of Multiple Myeloma: 30-Year Catamnesis

The historical association of defects in copper transport, increased serum copper levels, decreased ceruloplasmin levels, and copper deposition in the peripheral limbal cornea is well described in the clinical entity of Wilson (hepatolenticular) disease. However, a rarer pattern of copper deposition in the central cornea may occur with hypercupremia associated with monoclonal gammopathies. Copper deposition in the central Descemet membrane along with anterior and posterior lens capsules has been reported in individuals with multiple myeloma, benign monoclonal gammopathy, and IgG monoclonal gammopathy associated with pulmonary carcinoma. We report a 30-year catamnesis, a follow-up medical history of a patient who was first described in the ARCHIVES in 1975. She initially complained of blurred vision and a subjective change in her “iris color.” She was noted to have copper deposition in the Descemet membrane of the central cornea and in the anterior and posterior lens capsules and was subsequently diagnosed with hypercupremia secondary to a monoclonal gammopathy, an unusual variant of multiple myeloma.

Report of a Case. A 41-year-old white woman complained of slowly progressive, painless blurring of distance vision over the preceding several years. Her husband had commented on her changing “iris color” from blue to green. Her past medical history was unremarkable. She was taking no medications nor using alcohol or tobacco products. Her family history was negative for ocular disease.

Her best-corrected visual acuity was 20/20 OU at distance and Jager 0.5 OU at near. Intraocular pressures were normal. Biomicroscopy showed a pronounced iridescence sheen in the axial 7 to 8 mm of each cornea at the level of the Descemet membrane and a 2- to 3-mm circumferential limbal clear zone. The anterior and posterior lens capsules also had cocoa-powdery staining. The anterior vitreous was clear, and the fundus examination result was normal. Goldmann and tangent screen visual fields and electroneuroretinographic testing results were normal.

Color vision was evaluated with the Ishihara plates, Hardy-Rand-Rittler pseudosochromatic plates, the Farnsworth dichotomous panel D-15 test, the Farnsworth 100 hue test, and the Nagel anomaloscope.