Photorefractive Kerectomy for Correction of Epikeratophakia Regression

Excimer laser photorefractive kerectomy (PRK) is widely used for the correction of myopia, astigmatism, and hyperopia. It has also been used for correction of astigmatism after penetrating keratoplasty.

Epikeratophakia has been used in the treatment of nontolerant contact lens keratoconus patients. The epigrafts were made from machined corneal tissue that was found unsuitable for penetrating keratoplasty. Long-term follow-up of pediatric patients who underwent epikeratoplasty for optical correction of aphakia and were corrected for emmetropia revealed that later in life there is delayed myopic regression of the treated eye, which required further correction. In their patients, Colin et al failed to correct this myopic regression with PRK. We describe our experience with PRK for correction of delayed myopic regression of epikeratophakia in 4 eyes.

Design. All procedures were performed in the cornea and refractive surgery unit of The Goldschleger Eye Institute, Sheba Medical Center, Tel-Hashomer, Israel.

Four eyes of 2 twin sisters underwent epikeratoplasty at the ages of 8 and 9 years old because of very high myopia resulting from posterior lenticonus. Postsurgical refraction was stable for 8 years, then a rapid myopic regression of the epikeratophakic lenses was observed the following year (Table). Instead of removing the failed epikeratophakic lenses, we performed PRK on the eyes.

Results. Two and a half years after PRK, the refraction in all 4 eyes is stable and the epigrafts are clear. The Table presents the refraction and visual acuity results for the eyes before PRK and at 3 months, 1 year, and 2½ years after PRK. No haze has developed during this period. In all 4 eyes, a thin brown deposit ring was formed on the edge of the treated optical zone.

Comment. We describe herein our successful experience with PRK for regressed epikeratophakic lenses. After a follow-up of 2½ years, the results were stable and the epigrafts were clear. The eyes were also stable with regard to the best-corrected visual acuity.

Colin et al reported on 5 eyes with delayed refractive regression following myopic epikeratoplasty that were treated with PRK. Although the eyes were successfully corrected for emmetropia, all of them developed substantial subepithelial haze with poor visual acuity, and the epikeratophakic lenses had to be removed. It is possible that their poor results might be related to the preexisting corneal stromal abnormalities in their patients, which were not observed in our group. Thus, PRK can effectively be used to treat epikeratophakic regressed lenses in a selected group of patients in whom both the epikeratograft and the surrounding cornea are clear. This method eliminates the need for removal of the epikeratograft and exposing the patient to the risks of successive penetrating keratoplasty.

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Table

<table>
<thead>
<tr>
<th>Refraction, Degrees (Visual Acuity)</th>
<th>Right Eye</th>
<th>Left Eye</th>
<th>Right Eye</th>
<th>Left Eye</th>
</tr>
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<tbody>
<tr>
<td>Before epikeratoplasty</td>
<td>–12.50</td>
<td>–11.00</td>
<td>–16.00</td>
<td>–15.00</td>
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<tr>
<td>5 years after epikeratoplasty</td>
<td>–3.5 –2.0 × 30 (6/15p)</td>
<td>–3.5 –1.0 × 100 (6/20)</td>
<td>–5.0 –1.0 × 100 (6/15)</td>
<td>–5.0 –3.0 × 180 (6/12)</td>
</tr>
<tr>
<td>Before PRK</td>
<td>–17.00 (6/12)</td>
<td>–11.00 (6/12p)</td>
<td>–17.00 (6/12p)</td>
<td>–16.00 (6/12p)</td>
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<tr>
<td>3 months after PRK</td>
<td>–0.75 –2.0 × 15 (6/12+)</td>
<td>–3.0 –3.5 × 160 (6/15)</td>
<td>–3.75 –2.0 × 15 (6/12)</td>
<td>–4.0 –3.0 × 170 (6/15)</td>
</tr>
<tr>
<td>1 year after PRK</td>
<td>–1.0 –2.0 × 45 (6/12+)</td>
<td>–2.0 –3.0 × 135 (6/12)</td>
<td>–4.0 –2.0 × 25 (6/12p)</td>
<td>–5.0 –3.0 × 170 (6/12+)</td>
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<tr>
<td>2½ years after PRK</td>
<td>–3.00 (6/15+)</td>
<td>–2.5 –2.5 × 135 (6/12)</td>
<td>–3.00 (6/12p)</td>
<td>–4.0 –2.0 × 180 (6/12p)</td>
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*PRK indicates photorefractive kerectomy; p, partial. Boldface items indicate best-corrected visual acuity.


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Iatrogenic Keratoconus: Corneal Ectasia Following Laser In Situ Keratomileusis for Myopia

Laser in situ keratomileusis (LASIK) to correct myopia is performed by partially resecting a prescribed thickness of stroma, removing corneal tissue from the exposed stromal bed using the excimer laser, and then replacing the resected stromal tissue. This results in a substantial reduction of the biomechanically effective stress-bearing thickness of cornea provided by the residual stromal bed. There is concern that at some point, the tensile strength of the cornea might be reduced to the degree that progressive ectasia ensues, thereby resulting in steepening of the cornea, irregular astigmatism, and progressive myopia. This becomes a particularly contentious issue when, in the absence of classic clinical evidence of keratoconus, inferior steepening of the cornea seen on corneal topographic scan suggests the possibility of subclinical keratoconus. Herein, we report such a case of progressive ectasia following LASIK.

Report of a Case. A 23-year-old Hispanic man sought refractive surgery to correct myopia. Prior to surgery, he relied on spectacles to correct his vision. He reported infrequent changes in his prescription and good visual acuity in the years prior to consultation. His optometrist’s records prior to surgery documented a refraction of –12.75 – 2.25 × 65 OD and –8.50 – 1.50 × 79 OS, yielding a visual acuity of 20/30 OU. Findings from slitlamp examination (N.C.C.) revealed no characteristic corneal findings of keratoconus in either eye, including Vogt’s striae or a Fleischer ring. Ultrasound pachymetry measurements prior to surgery were 555 µm OD and 560 µm OS.

Corneal topographic scans of the right eye performed prior to surgery demonstrated mild inferonasal steepening with a maximum power of 44.5 diopters (D), simulated keratometry readings of 43.0 × 134/41.5 × 44, and a minimum keratometry reading of 41.3 × 37 (Figure 1, top). Keratoconus screening by Rabinowitz/McDonnell criteria suggested the presence of keratoconus, while screening by Klyce/Maeda criteria identified a 15% similarity to keratoconus. Corneal topographic scans of the left eye showed a homogeneously regular central corneal contour.

The patient underwent bilateral simultaneous LASIK (N.C.C.). A 130-µm flap was created using a manual microkeratome, and ablations 165 µm and 140 µm in depth were performed using an excimer laser (Summit Apex Plus; Summit Technologies, Waltham, Mass) in the stromal beds of the right and left eyes, respectively, estimated to leave residual stromal beds of 260 µm OD and 290 µm OS. On the first postoperative day, the patient’s uncorrected visual acuity was 20/40 OD and 20/60 OS, but at the next examination 6 weeks later, it had decreased to 20/400 OU, and his corrected visual acuity was 20/400 OD and 20/30 OS with a refraction of –5.50 sphere OU. A bilateral simultaneous enhancement procedure was performed under the flaps to fully correct the estimated residual myopia, and on the next examination 2 weeks later, the patient’s uncorrected visual acuity was 20/400 OD and 20/40 OS. Six weeks later, his best spectacle-corrected visual acuity was 20/25 OU and 20/30 OS, but at the next examination 6 weeks later, he reported infrequent changes in his prescription and good visual acuity in the years prior to consultation. His optometrist’s records prior to surgery documented a refraction of –12.75 – 2.25 × 65 OD and –8.50 – 1.50 × 79 OS, yielding a visual acuity of 20/30 OU. Findings from slitlamp examination (N.C.C.) revealed no characteristic corneal findings of keratoconus in either eye, including Vogt’s striae or a Fleischer ring. Ultrasound pachymetry measurements prior to surgery were 555 µm OD and 560 µm OS.

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Figure 1. Top, Preoperative corneal topographic map of the right eye demonstrating focal inferonasal steepening. Bottom, Preoperative corneal topographic map of the left eye demonstrating a homogeneously regular central corneal contour. Drop indicates diopters.
His best spectacle-corrected visual acuity decreased to 20/200 OD. Subjectively, the left eye remained relatively stable. On examination approximately 1 year following the initial procedures, his best spectacle-corrected visual acuity was 20/100 OD and 20/30 OS with a refraction of –10.50 – 4.00 × 67 OD and –3.75 sph OS. Findings from slitlamp examination revealed a hinged keratomileusis flap in both corneas and ectasia of the right cornea with a steep central protrusion. Ultrasound pachymetry measurements were 386 µm OD and 485 µm OS. A computed corneal topographic map of the right eye showed profound inferonasal steepening with an apical corneal power in excess of 50 D (Figure 2), while a map of the left eye showed a well-centered excimer laser ablation with marked flattening. The patient’s right eye was fitted with a rigid contact lens, with which he achieved satisfactory visual acuity.

Comment. Keratoconus is generally considered a contraindication for excimer laser refractive surgery, since it is expected that the progression of ectasia is likely to be hastened by the removal of central corneal tissue. Some authors have suggested that the risk is greatly exaggerated and have reported no evidence of acceleration of ectasia from 6 to 46 months after performing surface photorefractive keratectomy (PRK) on patients with a clinical diagnosis of keratoconus. While experience is limited and follow-up is brief, these results have prompted others to perform PRK in eyes that might be classified as forme fruste keratoconus; i.e., those eyes demonstrating topographic changes suggestive of keratoconus but without observable signs of keratoconus but in which corneal topographic scan findings are suspect. While neither eye demonstrated frank keratoconus, progressive ectasia occurred exclusively in the eye with a suspect corneal topography. Previous reports are few. Seiler and Quurke have also described progressive corneal ectasia that occurred in a patient with an asymmetric bow-tie pattern that they interpreted as forme fruste keratoconus. In our case, in addition to an asymmetric bow-tie pattern, other features suggested subclinical keratoconus, including markedly asymmetric topographic scan findings between the 2 eyes and poor initial best spectacle-corrected visual acuity.

It might be postulated that the risk of ectasia following LASIK might be higher than that following PRK because of the relatively thinner effective stress-bearing corneal stroma, but the residual stromal-bed thickness required to avoid progressive corneal ectasia in either topographically normal or abnormal eyes that undergo LASIK is unknown. Based
on personal experience, Barraquer\(^5\) has suggested a 300-µm thickness of stress-bearing cornea. By comparing the biomechanical properties of keratoconic corneas with normal corneas, Andreassen et al\(^6\) have estimated that for the normal cornea, a residual stromal-bed thickness of less than 250 µm might produce a cornea with a tangential elastic modulus comparable to that of a keratoconic cornea.

Lyle and Jin\(^7\) have reported a high incidence (26%) of progressive corneal ectasia that they termed *iatrogenic keratoconus* following hyperopic automated lamellar keratoplasty. The depth of the lamellar cut in this patient group ranged from 52% to 70%. However, their cases included corneas that had undergone prior radial keratotomy and were therefore structurally weakened to begin with. Hyperopic lamellar surgery frequently produces residual stromal beds less than 250-µm thick, but when performed in cases of primary hyperopia rather than consecutive hyperopia following radial keratotomy, progressive ectasia is relatively rare.\(^8\) In 3 cases of progressive corneal ectasia and keratoconus-like steepening developing in topographically normal-appearing eyes that underwent LASIK, Seiler et al\(^9\) estimated that the residual stromal-bed thickness was less than 200 µm in 1 patient and between 200 µm and 250 µm in the others. Based on this experience and on the theoretical calculations of Andreassen et al,\(^6\) these authors advocated a minimal residual stromal-bed thickness of 250 µm.

In the case we report, the estimated residual stromal-bed thickness prior to enhancement was 260 µm OD. This suggests that for certain corneas, such as those demonstrating features of keratoconus on topography, even 250 µm may not be an adequate stromal-bed thickness to prevent progressive ectasia. Alternatively, the actual thickness of the flap created by the microkeratome might differ substantially from the expected, which might produce a thinner than anticipated bed.\(^10\) Also, reading error in pachymetry measurements obtained, for example, from thicker paracentral regions of the cornea might also have led to overestimation of the residual stromal-bed thickness.

While these cases of corneal ectasia following LASIK have demonstrated fairly rapid progression, we are concerned that others may develop more slowly. In a study of loss of refractive effect in the first year following LASIK, Chayet et al\(^11\) report that regression seemed to be caused by an increase in corneal thickness rather than ectasia. However, longer-term studies are necessary. We strongly advocate that until we are better able to identify patients at risk for ectasia following LASIK, and the variables defining the biomechanical properties of the operated cornea are better described, LASIK should not be performed when findings from the examination or corneal topography suggest subclinical keratoconus.

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**Treatment of Conjunctival Mucosa-Associated Lymphoid Tissue Lymphoma With Intralesional Injection of Interferon Alfa-2b**

Conjunctival mucosa-associated lymphoid tissue (MALT) lymphomas are typically localized, low-grade tumors that differ histologically from other forms of primary extranodal non-Hodgkin lymphomas. Patients with conjunctival MALT lymphoma have been cared for with radiation, chemotherapy, surgical excision, cryotherapy, and even observation alone. To our knowledge, this is only the third report of conjunctival MALT lymphoma treated with local injection of interferon alfa-2b (IFN-α-2b)\(^1,2\) and the first to appear in the North American ophthalmic literature.

**Report of a Case.** A 21-year-old male student had a 2-month history of a painless, progressively enlarging mass on his right eye. Findings from anterior segment examination revealed a large salmon-colored lesion involving the right nasal conjunctiva, caruncle, and superior and inferior fornices (Figure 1). His uncorrected visual acuity was 20/20 OU and findings from the remainder of the ocular examination were unremarkable. The patient was tentatively diagnosed as having a conjunctival lymphoid neoplasm, pending biopsy and orbital imaging. He acknowledged a 40-lb weight loss during a 2-year period and moderate fatigue of recent onset. He denied having fever, chills, night sweats, other known masses, and risk factors associated with human immunodeficiency virus infection. He was otherwise healthy with no family history of hematological malignancy. Findings from physical examination demonstrated a thin man without evidence of lymphadenopathy or
hepatosplenomegaly. A conjunctival biopsy was performed. Sections of the conjunctival biopsy specimen showed a diffuse, small, cleaved lymphocytic infiltrate with occasional cells percolating into the epithelial layer (Figure 2). Immunoperoxidase stains were positive for CD20 (a B-cell marker) and negative for CD3 (a T-cell marker). Findings from flow cytometry demonstrated k light chain restriction (80%) and were positive for CD19 (a B-cell marker) (86%). A clonal rearrangement of the heavy chain, indicative of a clonal lymphoproliferative cell population, was demonstrated by polymerase chain reaction. No evidence of circulating lymphoma cells was found in either the blood or bone marrow by morphology, flow cytometry, or polymerase chain reaction. A computed tomographic scan of his head did not show orbital extension or intracranial abnormality. A computed tomographic scan of the chest, abdomen, and pelvis showed no abnormalities. We elected to treat this patient with an intralesional injection of 1 000 000 U of IFN-α-2b, administered 3 times weekly for a total of 12 doses. One month later, at the conclusion of treatment, the mass was considerably smaller. By 3 months, despite no additional therapeutic intervention, the conjunctival mass was undetectable (Figure 1). There is no evidence of recurrence after 3 additional months of follow-up, and the patient is otherwise healthy.

Comment. Lymphoid neoplasms of the conjunctiva are usually treated with radiotherapy. Bessell et al3 report no recurrence and a low rate of ocular morbidity in 115 patients receiving 30 or 40 Gy during 3 or 4 weeks; 8 (7%) developed radiation-induced cataracts, and 5 (4%) experienced disorders of ocular lubrication, despite shielding of the cornea and lens. Interferon α-2b is a glycoprotein produced by leukocytes possessing antiviral, immunoregulatory, and antitumor activity. The mechanisms responsible for its antitumor effects may include modulation of oncogenes and up-regulation of tumor cell surface antigens. As a single agent, its role seems limited to the treatment of low-grade lymphomas.4 Adverse effects include fevers, chills, and myalgias. Fatigue, anorexia, and weight loss may occur with protracted use. Complete remission was achieved in all 5 previously described patients with conjunctival lymphoma treated with local injection of IFN-α-2b.1,2 Adverse effects, if any, were not discussed. Our patient experienced transient headaches and nausea following the first 3 injections. The injection itself caused subconjunctival hemorrhage twice. The absence of vision-threatening complications and demonstrated efficacy, in this case, may warrant further consideration of the role of intrale- sional IFN-α-2b injection for treating conjunctival MALT lymphoma. Long-term follow-up of this and other cases is needed.

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Spontaneous Resolution of Vitreomacular Traction Documented by Optical Coherence Tomography

Optical coherence tomography (OCT) is an imaging technology that can clearly define the vitreoretinal interface. The preoperative and postoperative features of vitreomacular traction (VMT) have been described in a recent case report. In the present case, serial OCT images documented the natural history of VMT secondary to intermediate uveitis. This is the first report demonstrating spontaneous resolution of VMT with OCT.

Report of a Case. The patient is a 60-year-old white woman with a 1½-year history of stable, bilateral intermediate uveitis treated with prednisolone acetate drops twice daily in both eyes. Five months before her initial visit to us, her visual acuity dropped to 20/60 OS from 20/25 OS. Six weeks before referral to us, visual acuity dropped further to 20/80 OS and a sub-Tenon triamcinolone acetonide injection was administered in the left eye. The patient was referred to our retina service for evaluation of continuing decreased vision in her left eye.

Best-corrected Snellen visual acuity was 20/30 OD and 20/70 OS. Applanation tonometry revealed intraocular pressures of 19 mm Hg in the right eye and 27 mm Hg in the left, likely secondary to the corticosteroid injection. Slitlamp examination revealed trace pigmented cells in the anterior chambers of both eyes. Dilated fundus examination revealed persistent vitreous traction at the fovea although retinal thickening persisted in the setting of quiescent uveitis.

One month later the patient reported spontaneous improvement in vision. Best-corrected visual acuity improved to 20/40 OS. Retinal examination revealed persistent vitreous traction at the fovea although retinal thickening appeared to be decreased. The OCT image of the left eye revealed that the posterior hyaloid had largely detached from the retina although a focal area of adhesion...
remained temporal to the fovea (Figure, B).

Four months after the initial visit the patient reported no visual complaints and visual acuity improved to 20/30 OS. Retinal examination revealed resolution of the central retinal thickening and a taut posterior hyaloid anterior to the fovea. The OCT image confirmed complete detachment of the posterior hyaloid, diminished retinal thickening, and restoration of the normal foveal contour (Figure, C).

Comment. Vitreomacular traction is a result of detachment of the posterior vitreous with persistent vitreomacular adhesions that produce retinal thickening and cystic changes.2 In this case the cause appears to have been intermediate uveitis because the pattern of adhesion and the patient’s course after vitreomacular separation are consistent with previous descriptions by Schepens et al.2

Optical coherence tomography is a diagnostic technique that may be useful in distinguishing the morphologic characteristics of a wide variety of retinal abnormalities.4 This case illustrates the utility of OCT in diagnosis and follow-up of patients with VMT. Over a 4-month period, our patient’s condition spontaneously improved, both subjectively and on the OCT images, and this is attributed to detachment of the posterior hyaloid from the macular surface. As spontaneous resolution may occur in some eyes with VMT, a period of observation with sequential OCT evaluations to assess the vitreous anatomy may be considered prior to surgical intervention.

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Acute Bilateral Visual Loss Associated With Retinal Hemorrhages Following Epiduroscopy

Acute bilateral visual loss associated with retinal hemorrhages following epidural steroid injection or gas myelography has been described.1,3 To our knowledge, we report the first case of acute bilateral visual loss associated with preretinal, retinal, and subretinal hemorrhages occurring after epiduroscopy, a diagnostic and therapeutic procedure that allows visualization of the spinal cord and epidural space in patients with chronic back pain.

Report of a Case. An 80-year-old woman whose medical history was significant only for chronic back pain and bilateral lower extremity weakness unresponsive to medical management underwent diagnostic epiduroscopy. The patient was brought to the operating room and placed in the prone position. Needle placement in the epidural space was confirmed with fluoroscopic guidance in 3 views. A guidewire was inserted and the scope apparatus was introduced. Visualization of the epidural space was allowed via saline instillation at a pressure not exceeding 60 mm Hg for a period of less than 5 minutes. Epiduroscopy was performed without complication under intravenous sedation; the patient’s vital signs remained stable throughout the procedure. Immediately following the procedure, the patient noted acute visual loss in both eyes.

Ocular examination at that time revealed a best-corrected visual acuity of 5/200 OD and 3/200 OS. Intraocular pressures and pupils were within normal limits. Anterior segment examination was notable for well-positioned intraocular lenses. Dilated funduscopic examination was remarkable for multiple, round, preretinal, retinal, and subretinal hemorrhages involving the posterior pole and midperiphery in both eyes (Figure 1). The disc margins were sharp and the retinal vessels were of normal caliber. Fluorescence angiography revealed leakage of dye from the subretinal hemorrhages in both eyes. The patient was taken to the operating room for vitrectomy with peeling of preretinal membranes under general anesthesia. A total of 21 months after epiduroscopy, the patient’s vision had improved to 20/200 OD and 20/70 OS. The retinal hemorrhages had resolved, although subretinal fluid persisted in both eyes.

Figure 1. Color fundus photographs of the right (left) and left (right) eyes demonstrate multiple retinal hemorrhages involving the posterior pole.
cein angiography revealed areas of blocked fluorescence corresponding to the hemorrhages seen clinically (Figure 2).

Six months later, best-corrected visual acuity had improved to 20/100 OD and 20/80 OS. The retinal hemorrhages had resolved spontaneously in both eyes (Figure 3); vision was limited secondary to non-exudative age-related macular degeneration.

Comment. The occurrence of retinal hemorrhages in one or both eyes immediately after the injection of oxygen into the subarachnoid space during myelography or following epidural injection of corticosteroids has been described previously. To our knowledge, this is the first report of bilateral retinal hemorrhages occurring after epiduroscopy, a procedure involving placement of a fiberoptic scope into the epidural space with injection of saline for visualization of the spinal cord and subarachnoid space.

Increased cerebrospinal fluid (CSF) pressure during epiduroscopy may lead to retinal hemorrhages by 2 possible mechanisms. First, increased CSF pressure may be transmitted directly through the optic nerve sheaths to the retinal venous circulation. Alternatively, increased CSF pressure may result in decreased cerebral blood flow, which in turn stimulates a reflex increase in ophthalmic artery pressure with resultant venous collapse and rupture of capillaries.

Acute visual loss associated with retinal hemorrhages is an uncommon but significant complication of epiduroscopy. Experimental evidence suggests that elevation of CSF pressure may be modulated by decreasing the rate and volume of epidural injection, potentially lowering the likelihood of this ocular complication. Patient education and informed consent are paramount when recommending this procedure, especially for patients with bleeding tendencies or compromised retinal circulation.

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Capillary Hemangioma of the Optic Nerve Head and Juxtapapillary Retina

A 24-year-old man was noted to have decreased vision in the left eye on a routine eye examination. Funduscopy revealed a mass overlying the optic nerve head and adjacent superotemporal retina in the left eye, as well as a large amount of exudate in the macula. Fluorescein angiography demonstrated early hyperfluorescence of the lesion and late pooling of the dye, outlining the retinal detachment. A diagnosis of capillary hemangioma of the optic nerve head and juxtapapillary retina was made. The lesion was treated with argon laser photocoagulation. Subsequent progression of the tumor led to a total retinal detachment and rubeosis iridis and resulted in enucleation. Histopathologic examination of the lesion revealed a mass composed of fine capillaries, lined with endothelium, and filled with red blood cells. Capillary hemangioma of the optic nerve head and juxtapapillary retina is rare, and only a few reports of its histopathologic characteristics exist. The association of the tumor with von Hippel-Lindau syndrome should be considered by ophthalmologists.

Retinal capillary hemangioma is a rare, benign vascular tumor of the retina. Two variants of the tumor are recognized: peripheral retinal capillary hemangioma and the less frequent capillary hemangioma of the optic nerve head and/or juxtapapillary retina. Either variant can be associated with von Hippel-Lindau syndrome. Retinal capillary hemangioma usually occurs in the second or third decade of life with equal frequency in both sexes and no racial predilection. Hereditary and nonhereditary forms exist. Initial symptoms include decreased visual acuity and visual field loss. The lesion may be asymptomatic and discovered on a routine ophthalmic examination. An untreated tumor frequently enlarges, and, over time, leads to progressive subretinal and intraretinal exudation and retinal detachment. We present a case of a capillary hemangioma of the optic nerve head and juxtapapillary retina in a 24-year-old man.

Report of a Case. A 24-year-old white man was noted to have decreased visual acuity in the left eye on a routine eye examination. He had not been aware of the visual loss. His medical history was unremarkable. Visual acuity was 20/20 OD and counting fingers at 1.2 m (4 ft) in the left eye. On dilation of the pupil, a discrete, elevated, reddish mass obscuring the optic nerve head was noted in the left eye (Figure 1). The elevated portion of the mass was approximately 4.5 mm in diameter. Contact B-scan ultrasonography demonstrated the mass measured 5.4 mm thick and 11.5 mm × 9.4 mm in diameter at the base. Dilated, tortuous feeder vessels, arising from the optic disc, were seen on the surface of the lesion. Subretinal lipid exudate surrounded the lesion, and an exudative retinal detachment involving the macula was present. No other hemangiomatous lesions were present in the left eye. Results of the fundus examination of the right eye were normal. Fluorescein angiography demonstrated early hyperfluorescence of the vascular lesion and late pooling of the dye, outlining the retinal detachment (Figure 2).

Based on the clinical appearance of the mass and the supporting diagnostic studies, a diagnosis of capillary hemangioma of the optic nerve head and juxtapapillary retina was made. The tumor was treated with argon laser photocoagulation on 2 occasions. Following this treatment, the patient was lost to follow-up for 8 months. Upon his return to care, he had developed a total retinal detachment and rubeosis iridis, and the eye was enucleated. Although the patient had no family history or symptoms suggesting von Hippel-Lindau syndrome, he was advised to undergo testing to rule out cerebellar hemangioblastoma or other visceral tumors, but he failed to comply with that recommendation.

The gross section of the enucleated eye revealed a funnel-shaped retinal detachment with xanthochromic subretinal fluid containing refractile particles and a white mass lesion (Figure 3). The mass, measuring 4.0 mm wide and 3.0 mm thick, arose mainly from the optic nerve head and involved the inner retina superotemporally. Histopathologic examination demonstrated a total retinal detachment with a massive amount of subretinal fluid with numerous cholesterol clefts, corresponding to the refractile particles seen on a gross section (Figure 4). A lobulated mass composed of fine capillaries lined with normal endothelium and filled with red blood cells was seen. The vascular chan-

Figure 1. Fundus photograph of the left eye showing a discrete, elevated, reddish lesion obscuring the optic nerve head. Note the surrounding subretinal lipid exudate.

Figure 2. Contact B-scan ultrasonography demonstrated the mass measured 5.4 mm thick and 11.5 mm × 9.4 mm in diameter at the base. Dilated, tortuous feeder vessels, arising from the optic disc, were seen on the surface of the lesion. Subretinal lipid exudate surrounded the lesion, and an exudative retinal detachment involving the macula was present. No other hemangiomatous lesions were present in the left eye. Results of the fundus examination of the right eye were normal. Fluorescein angiography demonstrated early hyperfluorescence of the vascular lesion and late pooling of the dye, outlining the retinal detachment (Figure 2).
nels were separated by vacuolated interstitial cells (Figure 5). In the anterior segment of the eye, rubeosis iridis and anterior displacement of the iris and lens were apparent.

Comment. Capillary hemangioma of the retina most likely represents a vascular hamartoma and was first described by von Hippel in 1904. Lindau reported the association between retinal capillary hemangiomas and central nervous system tumors. The term “von Hippel-Lindau syndrome” is applied to an autosomal dominant disorder, recently linked to abnormalities in the short arm of chromosome 3 (locus 3p25), characterized by cerebellar and spinal cord hemangioblastomas, renal cell carcinoma, pheochromocytoma, and retinal angiomas.

Peripheral capillary hemangioma of the retina is often multifocal and bilateral and has the characteristic appearance of a reddish or gray nodule in the peripheral retina supplied by enlarged, tortuous feeder vessels. The well-circumscribed endophytic capillary hemangioma arising from the optic nerve head can also be recognized clinically without difficulty. When sessile or exophytic, capillary hemangioma of the juxtapapillary retina and optic nerve head may be misdiagnosed as unilateral papilledema, papillitis, choroiditis, choroidal neovascularization, or choroidal hemangioma. Stereoscopic fluorescein angiography is often helpful in establishing the diagnosis of retinal angioma. Fluorescein angiography shows hyperfluorescence of the vascular lesion in the middle and later retinal stages, often followed by continuous leakage from the lesion in the later stages of the study. Dilated feeder vessels can be readily visualized with angiography.

Histologically, the tumor is composed of numerous capillaries lined by normal endothelium, separated by plump, vacuolated interstitial cells, containing lipidlike material. Juxtapapillary capillary angioma has been shown histologically to have a choroidal as well as a retinal blood supply. With time, the capillaries within the hemangioma become incompetent, which presumably leads to progressive subretinal and intraretinal exudation in the macula, often resulting in a total retinal detachment. Spontaneous regression of a retinal hemangioma has been described, but this is a rare occurrence.

Eyes with untreated retinal angiomas tend to have a poor prognosis because of the associated macular exudate and exudative retinal detachment. Early treatment of peripheral capillary hemangioma lesions with argon and xenon photoacoagulation is advocated and tends to be more successful with tumor size of 1 disc diameter or smaller. Multiple treatment sessions with...
moderate-intensity, prolonged duration photocoagulation are recommended. Other treatment modalities such as cryotherapy, diathermy, and proton beam irradiation have been used as well. Photocoagulation of juxtapapillary lesions is associated with poor outcomes, since destruction of nerve fiber layer in the macula during photocoagulation may contribute to the visual loss. Therefore, treatment of juxtapapillary angiomas is generally not recommended until macular exudation threatens the central vision. It is also unlikely for photocoagulation to penetrate a large endophytic lesion, such as the one described in this case report.

Retinal capillary hemangiomas is often associated with von Hippel-Lindau syndrome, but the actual incidence is not precisely known and may have been underestimated in previous studies. Schindler et al. found a 24% incidence of von Hippel-Lindau syndrome or a positive family history in 55 patients with capillary angioma of the optic disc. Thus, it is important to obtain a family history and refer the affected individual for the appropriate systemic studies, which include magnetic resonance imaging of the brain and spinal cord to rule out hemangioblastomas, and abdominal computed tomography to rule out renal and other visceral cysts. Urinary studies for pheochromocytoma have also been advocated. The patient’s relatives should undergo a dilated fundus examination to exclude retinal angiomas.

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Sildenafil (Viagra) Associated Anterior Ischemic Optic Neuropathy

Sildenafil citrate (Viagra) has been shown to be effective for erectile dysfunction and is well tolerated. Headache and flushing are common side effects. We describe the novel finding of ischemic optic neuropathy in a patient concomitant with his ingesting sildenafil.

Report of a Case. A 52-year-old healthy man with no known vasculopathic risk factors suffered from erectile dysfunction after transurethral resection for prostate cancer. He took his first dose of 50 mg of sildenafil citrate in the evening and, within 1 hour, sweating and a severe generalized headache developed. He saw blue “lightning bolts” and reported blurry vision in both eyes. This lasted 30 minutes, but the vision in the left eye remained blurred inferiorly. No erection occurred and he did not have sex. He tried the medication the next night with recurrence of the same symptoms. The blurry vision of the left eye did not change. He had Crohn disease and took methylphenidate hydrochloride for attention deficit disorder in the morning and at noon. He was not a headache sufferer.

Ophthalmic examination 5 days later showed corrected visual acuities of 20/20 OU and normal color vision (Ishihara test) in both eyes. There was no relative afferent pupillary defect. Kinetic perimetry revealed an inferior altitudinal visual field depression in the left eye (Figure 1). Dilated funduscoppy showed superior swelling of the left optic nerve head and a normal retina consistent with an ischemic optic neuropathy (Figure 2). The right eye had a normal optic nerve with a 0.1 cup-disc ratio. Findings from the remainder of the ophthalmic examination were normal; results of a thorough laboratory evaluation were unrevealing. Ophthalmic examination 9 months later showed pallor superiorly in the left eye.

Comment. Transient changes in perception of color hue or brightness have been reported with sildenafil. Electrophysiological reductions on electroretinograms have also been described although all patients were clinically asymptomatic. The exact mechanism for these changes is unclear. However, the retina has a higher concentration of phosphodiesterase type 6 and the visual phenomena may be due to the ability of sildenafil to weakly block this enzyme.

The only neuro-ophthalmologic complication reported thus far is a third nerve palsy. This patient developed diplopia 36 hours after ingestion of 50 mg of sildenafil citrate. The patient smoked and took amitriptyline therapy. It was unknown if the patient developed an erection or successfully completed intercourse. Donahue and Taylor hypothesized that sildenafil caused a hypotensive ischemic event to the third nerve. This is supported by the ability of sildenafil to potentiate the hypotensive effects of nitrates. Their patient, however, developed his symptoms after a significant period. We believe this raises some doubt concerning the causation of the oculomotor palsy by sildenafil therapy. In contrast, our patient developed symptoms within 1 hour of ingesting sildenafil. It is unlikely that our patient’s event was complicated by
methylphenidate therapy because he was not taking the long-acting, sustained-release formulation and sildenafil was ingested many hours later. He also did not develop an erection or participate in any sexual activity and thus no steal phenomenon can be postulated as the cause of the probable hypotensive event to his optic nerve.

Our patient had a very small cup-disc ratio. Sildenafil therapy may have potentiated the same mechanisms that theoretically induce ischemic optic neuropathy in an anatomically susceptible optic nerve head, specifically hypotension. Although it cannot be explicitly proven that sildenafil therapy was the cause of this patient’s optic neuropathy, the close temporal relationship strongly suggests this.

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