Corneal Findings in Hemochromatosis

Corneal pigmentation deposition has been reported in many systemic diseases. These include the lysosomal diseases, Wilson disease, amyloidosis, multiple myeloma, cystinosis, and hemochromatosis. These underlying conditions must be considered in the differential diagnosis of corneal deposits, as they might be treatable. We focus on an unusual case of corneal iron deposition in a patient with acquired hemochromatosis.

Report of a Case. A 70-year-old Russian woman immigrated to the United States in 1995. She was seen by the ophthalmology department because of gradually worsening vision since 1990. Her medical history was notable for type II diabetes, hypertension, arthritis, anemia, gastritis, and liver dysfunction. She had been diagnosed with anemia in 1988, and received weekly intravenous iron injections between 1988 and 1989 after daily injections for 3 weeks. The dosage of each injection and the type of anemia that was present could not be ascertained. She denied alcohol abuse, previous ocular trauma, or the use of any topical ophthalmic medications. The patient received multiple phlebotomy treatments in 1997.

At the time of our initial ophthalmic examination in March 1996, the best-corrected visual acuity was 20/40 OD and 20/40 OS. The patient's eyelids were not abnormal in color; the lenses had a mild degree of nuclear sclerosis. The conjunctiva and cornea of both eyes showed small fine brown pigmented deposits. The diffuse corneal pigment was slightly greater inferiorly than superiorly, with a clear zone of approximately 1.5 mm between the pigment and the limbus, in the intraepithelial and anterior third of the stroma (Figure 1 and Figure 2). The conjunctival pigmentation was extremely fine, diffuse, and interpalpebral. The extremities revealed no abnormal pigmentation.

Laboratory test results since 1995 have revealed serum ferritin levels between 793 and 1173 µg/L (reference range, 10-291 µg/L), with a normal iron-binding capacity, and abnormal liver function. A liver biopsy specimen in 1996 revealed iron pigment in parenchymal cells and cirrhotic changes consistent with hemochromatosis.

Comment. Hemochromatosis can be genetic (primary) or acquired. In genetic hemochromatosis, intestinal iron absorption is significantly increased, and causes deposition of iron in the liver, skin, pancreas, joints, and heart. The organs involved become impaired with continued accumulation.

Acquired hemochromatosis occurs from hemolysis (usually from thalassemia or sideroblastic anemia) or multiple transfusions. Unlike primary hemochromatosis, iron deposition occurs first in reticuloendothelial cells. As the process continues, the reticuloendothelial system becomes saturated, and parenchymal cell deposition also occurs, leading to many of the same systemic manifestations as genetic hemochromatosis.

Ocular hemosiderosis is iron toxicity confined to the eye due to a retained intraocular iron foreign body or persistent intraocular hemorrhage. The iron deposition occurs in most parts of the eye, but especially involves the posterior segment. Severe visual loss can result from vitreal degeneration with formation of contraction bands and retinal degeneration.

The ophthalmic manifestations of genetic or acquired hemochromatosis are quite different from the effects of ocular hemosiderosis. In genetic or acquired
disease, the deposition is generally limited to the sclera and ciliary body.\textsuperscript{1,2} Hudson\textsuperscript{3} analyzed 5 post-mortem cases of biopsy-proven hemochromatosis and observed no abnormalities other than conjunctival aneurysms and fundus changes similar to early diabetic retinopathy (2 of the 5 patients had diabetes). In fact, Hudson stated that “no other ocular findings characteristic of hemochromatosis have been observed.”\textsuperscript{3}

Our patient is, to our knowledge, the first reported case of corneal iron deposition in acquired hemochromatosis. Only 1 previous report in the literature has described a similar clinical picture. Urrets-Zavalia and Katz\textsuperscript{2} noted corneal iron deposition and termed it “corneal hemochromatosis,” because there was no systemic iron overload. They described a “multitude of minute refringent dots in the anterior third of the corneal stroma,” an appearance similar to our patient. Both eyes required penetrating keratoplasty due to corneal opacity. Davies et al\textsuperscript{2} reported conjunctival pigmentation encroaching onto the limbus in their series of patients with hemochromatosis. No treatment was advised for our patient, as her visual acuity was good. Furthermore, as it may require 2 to 3 years for phlebotomy to reduce substantially the total body iron stores, the intracorneal iron theoretically may also clear with the passage of time.

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Endophthalmitis Induced by Chryseomonas indologen

Bacterial endophthalmitis is a rare ocular disease that often has a poor visual outcome. The investigators for the Endophthalmitis Vitrectomy Study have developed guidelines for treatment of patients with endophthalmitis in addition to reporting prognostic indicators. Although visual acuity at the time of evaluation was the best predictor of final visual outcome, microbiological factors seem to play an important predictive role. Patients with endophthalmitis related to gram-negative organisms fared considerably worse compared with cases associated with gram-positive organisms.

We report a case of gram-negative endophthalmitis following cataract surgery induced by Chryseomonas indologen. To our knowledge, this description represents the first reported case of endophthalmitis caused by this organism.

Report of a Case. A healthy 92-year-old white man underwent phacoemulsification of a cataract of the left eye, during which a dehiscence of the posterior capsule and posterior displacement of lens material occurred. An anterior vitrectomy was performed and a posterior chamber intraocular lens was placed in the ciliary sulcus. During rotation of the implant, it subluxed into the vitreous cavity. An anterior chamber intraocular lens was then inserted.

Though the patient experienced persistent intense pain and redness of the left eye that began the night of the surgery, he did not return for follow-up examination until 7 days later, when he was referred to the University of British Columbia Eye Care Center, Vancouver.

Examination disclosed a visual acuity of 20/30 OD and hand motions OS. The intraocular pressures were 20 mm Hg OD and 45 mm Hg OS. Slitlamp examination disclosed a markedly edematous left cornea with cells (+4+) and flare in the anterior chamber with extensive fibrin and a layered hypopyon (Figure). Fundus examination revealed only a hazy red reflex.

The patient underwent an immediate anterior chamber and vitreous aspiration followed by an intravitreal injection of 1 mg of vancomycin (1 mg/0.1 mL), 2 mg of ceftazidime (2 mg/0.1 mL), and 1 mg of dexamethasone (1 mg/0.1 mL). Therapy was begun with acetazolamide, 250 mg once daily; 1% prednisolone acetate once daily; 0.5% timolol maleate twice daily; and ciprofloxacin drops hourly. Six hours later, the pain had largely subsided. The clinical appearance remained unchanged except for a reduction of the intraocular pressure to 25 mm Hg OS.

The vitreous specimen revealed gram-negative rods and polymorphonuclear cells (2+). The aqueous specimen contained polymorphonuclear cells (1+) but no organisms. Microbiological cultures subsequently grew Chryseomonas indologen from both specimens.

One month later, the visual acuity had improved to 20/200 OS. The fibrin in the anterior chamber had largely resolved. The peripheral retina was visible but the presence of vitreous debris obscured a clear view of the macular region. No retinal necrosis was present. Ten weeks after the antibiotic injection, the visual acuity was 20/70 OS. Mild persistent corneal edema was noted. Results of a fundus examination were unremarkable.

Comment. Chryseomonas indologen is an aerobic motile gram-negative rod with a distinct yellow-orange pigment seen most commonly in petroleum-contaminated soils. Chryseomonas is one of a few bacteria species that is capable of utilizing hydrocarbons as its sole source of carbon and energy. This group of organisms rarely causes systemic infections, and such cases are often related to the introduction of indwelling lines or prostheses. It morphologically resembles Pseudomonas species and was originally categorized in that genus. The 2 organisms are phylogenetically similar and share select common RNA sequences.
Pars plana vitrectomy was performed in the left eye in July 1992. Histopathologic stains were consistent with amyloidosis. The patient’s postoperative visual acuity improved to 20/20 OS, but amyloidosis has a high recurrence rate. Vitrectomy in the right eye was performed in August 1992, with concurrent endolaser panretinal photocoagulation to destroy the possible site of production or secretion. In May 1993, the patient was symptom free with visual acuity of 20/20 OU; however, results of her ophthalmic examination showed asymmetric intraocular pressure of 16 mm Hg OD and 21 mm Hg OS, with a clear posterior segment in the right eye and opacities in the vitreous in the left eye. In June 1994, visual acuity had deteriorated to 20/25 OD and 20/80 OS, with intraocular pressure at 14 mm Hg OD and 57 mm Hg OS. The left eye showed an afferent pupillary defect, large recurrent vitreous opacities, optic nerve cupping, and marked visual field loss.

Maximum medical treatment did not sufficiently control intraocular pressure, and a mitomycin-C augmented trabeculectomy failed after several months. In February 1995, the patient underwent pars plana vitrectomy with endolaser panretinal photocoagulation and placement of an Ahmed glaucoma drainage tube. The patient has done well since that time; the most recent ophthalmic examination in July 1997 showed a visual acuity of 20/30 OD and 20/40 OS consistent with mild cataract, intraocular pressure of 10 mm Hg OD and 13 mm Hg OS, and clear vitreous in both eyes. She has been diagnosed with cardiac, neurological, and gastrointestinal deposition of amyloid.

Comment. Amyloidosis is considered a complex of related diseases characterized by extracellular deposition of amyloid protein. Amyloid fibrils are composed of various proteins arranged in a β-pleated sheet configuration. Various protein abnormalities can cause amyloidosis. Genetically aberrant prealbumin is thought to be responsible for all cases of vitreoretinal amyloidosis in which the deposits have been chemically sequenced. Prealbumin has a high proportion of proteins in the β-pleated sheet structure, which may polymerize into the amyloid fibril. Prealbumin synthesis has been seen in retinal pigment epithelium, although the major source is considered to be the liver. Other research suggests that vitreoretinal amyloid has a retinal vascular origin.

The mechanism by which laser panretinal photocoagulation led to resolution of vitreoretinal amyloidosis is unclear. A secondary effect on the retinal vasculature could have reduced secretion of amyloid formed elsewhere, eg, in the liver. Alternatively, destruction of retinal pigment epithelium could have destroyed the site of amyloid synthesis. In our patient, glaucoma was probably due to amyloid obstructing trabecular meshwork. The panretinal argon laser photocoagulation probably helped reduce the amount of amyloid deposited in the trabecular meshwork.

Panretinal argon laser photocoagulation prevented recurrence of amyloid deposits in the vitreous. The small number of individuals with ocular amyloidosis prevents investigating laser therapy with a clinical trial. We are optimistic, however, that our observations may prove useful in treating other patients with this condition.

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Pulmonary Hypertension and Diffuse Macular Edema Responsive to Acetazolamide

We report a case of primary pulmonary hypertension (PPH) that led to uveal effusion and macular edema. In our patient, these symptoms rapidly resolved with the administration of acetazolamide.

Report of a Case. A 33-year-old woman had a 3-month history of bilateral blurred vision and eye pain. She was treated with 30 mg of prednisolone daily (0.5 mg/kg) for presumed posterior scleritis for 3 weeks with no improvement and was referred to our unit. A diagnosis of PPH had previously been made on the basis of right-sided heart failure with an elevated pulmonary arterial pressure of no obvious cause. Maintenance therapy consisted of diuretics, vasodilators (diltiazem hydrochloride), and anticoagulation. Her mother also had PPH.

On examination, corrected visual acuity was 20/90 OD and 20/40 OS. There was bilateral conjunctival chemosis with dilated nonarterialized episcleral vessels, but no anterior uveitis. Intraocular pressures were 10 mm Hg OD and 9 mm Hg OS. Fundus examination revealed bilateral macular edema with retinal pigment epithelium mottling (Figure 1, left) and bilateral inferior serous retinal detachments. On general examination, blood pressure was 110/70 mm Hg and signs of right-sided heart strain including elevated jugular venous pressure, ankle edema, right ventricular heave, systolic murmur, and a palpable liver edge were evident.

B-scan ultrasonography demonstrated choroidal thickening and fluorescein angiography demonstrated widespread retinal pigment epithelium mottling and macular edema without retinal vascular leakage (Figure 1, right). A complete blood cell count showed a hemoglobin level of 165 g/L (reference range, 120-150 g/L) and a hematocrit of 0.49 (reference range, 0.36-0.47) in keeping with the polycythemia of chronic hypoxia. Results of routine biochemistry testing were normal.

Treatment with oral acetazolamide (sustained release, 250 mg twice daily) was begun, with resolution of the serous retinal detachments and macular edema within 6 days, resulting in unaided visual acuities of 20/20 OD and 20/15 OS. Systemic symptoms and signs were unchanged.

Subsequent discontinuation of treatment after 6 months resulted in recurrence of symptoms and signs in the left eye within 1 week. Optical coherence tomographic scans were performed and demonstrated edema and serous elevation of the macula (Figure 2, left). Oral acetazolamide was reintroduced and optical coherence tomographic scanning after 23 hours demonstrated an almost complete resolution of both (Figure 2, right).

Comment. Primary pulmonary hypertension is a rare condition and is often fatal due to progressive right-sided heart failure with vasoconstriction, vascular wall remodeling, and thrombosis in situ of small pulmonary arteries. Its cause is unknown, but the gene for familial PPH (a minority of cases) has been recently localized to chromosome 2q31-32.

Bilateral serous detachments similar to the uveal effusion syndrome have been reported before in PPH and variously attributed to retinal and choroidal stasis, oxygen-induced vasoconstriction, elevated venous pressure and Valsalva effect due to vomiting, but acetazolamide has not been previously tried in these patients. Elevated venous pressure in the orbit could lead to this clinical picture as, for example, uveal effusion has been reported in carotidocavernous fistulae. However, right-sided heart failure in the more common secondary pulmonary hypertension has not been reported to cause macular...
edema and serous detachment. The remarkable feature in our patient was the appearance of diffuse macular edema without retinal vascular leakage. Its rapid disappearance with acetazolamide, a drug known to resolve macular edema in other disorders of retinal pigment epithelium, suggests retinal pigment epithelial dysfunction in our patient.

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Traumatic Foveal Relocation With Good Visual Acuity

Neovascular age-related macular degeneration is a major cause of visual loss in elderly patients.1 Treatment options are limited for most of these patients. Surgical relocation of the foveal retina to an area of healthy retinal pigment epithelium (RPE) has been described.2-6 We report a case of traumatic foveal relocation with good visual acuity.

Report of a Case. An 18-year-old man suffered a bottle rocket injury with resultant trauma to his left eye. His right eye was not injured and had an uncorrected visual acuity of 20/20. Left eye evaluation revealed light perception visual acuity without associated light projection. A 50% hyphema and corneoscleral laceration were present. The laceration extended 2 mm posterior to the limbus at the 9- and 12-o’clock positions, involved the superonasal corneal quadrant, and extended to the center of the visual axis. Repair of the laceration with excision of prolapsed iris was performed the day of the injury by the referring physician.

The patient was seen at our institution 4 days following his injury. Left eye visual acuity was light perception, intraocular pressure was 10 mm Hg, and an ophthalmic examination showed an intact corneal wound. Corneal edema, disrupted lens material, and a hemorrhage precluded a view to the retina. Ultrasonography revealed a possible superonasal retinal detachment.

A vitrectomy was performed 18 days after his injury, removing the traumatically disrupted lens and hemorrhagic vitreous. The corneal clarity had improved adequately to proceed without a temporary keratoprosthesis.

The retina was tractionally elevated in a ridge extending from the optic disc to the 11-o’clock position periphery where depressed examination revealed retinal incarceration in the original rupture site. The borders of the ridge had a concave appearance and no evidence of a break was found. The macula was attached, but rotated superiorly. The vitreous base was trimmed 360° with scleral indentation and a 3.5-mm circumferential buckle was placed to support the vitreous base. No gas or silicone oil was used.

The subretinal fluid beneath the tractional retinal elevation resolved, resulting in a dry fold postoperatively (Figure 1). His remaining retina remained attached. His fovea was found to be rotated superiorly 43° with the darker RPE of the central macula visible beneath the retina adjacent to the inferotemporal arcade vessels. Fine spiral folds were seen at the supertemporal disc margin.
(Figure 2). He fixated reliably on the area of foveal relocation and had a corrected visual acuity of 20/60 OS at 6 weeks. The visual acuity was obtained despite vascularization of the corneal wound and corneal sutures within the visual axis. He reported excyclorotation of images in his left eye.

Comment. Therapy for neovascular age-related macular degeneration is limited. Photocoagulation of choroidal neovascularization is useful for a small percentage of patients. Surgical removal of choroidal neovascularization has limited success in improving vision.1

In age-related macular degeneration many components of the complex of macular RPE, Bruch’s membrane, and choriocapillaris are dysfunctional. Surgically relocating the fovea to an area of healthy RPE that may preserve foveal function has been proposed. Different surgical methods have been described to accomplish foveal relocation.2-4

The macular area is characterized by the greatest density of RPE melanin pigmentation5 and a lobular choroidal angioarchitecture that allows for extremely fast circulation.6 The ability of extramacular RPE and choriocapillaris to support good foveal function is relatively unknown.

Our patient demonstrates good visual acuity following foveal relocation to an area of extramacular RPE. Assuming comparatively good extramacular RPE function in patients with age-related macular degeneration, foveal relocation may offer a surgical alternative to the limited treatment options available to those with subfoveal choroidal neovascularization.

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Eyelid Neuroma Associated With Swim Goggle Use

The use of plastic swim goggles has been associated with several complications including supraorbital neuralgia, periorbital leukoderma, contact dermatitis, and even traumatic ruptured globe.1,2 Others have reported on the “competition swimmer’s eyelid syndrome,” a pseudo-baggy eyelid localized to the medial part of the superior eyelid.3 The authors described 2 young, competitive swimmers with localized swelling of the upper eyelid related to their use of swim goggles. The report did not include microscopic analysis of the involved tissue, and the authors believed that the swelling was due to microtrauma from the rims of the plastic goggles.

We report our experience with the swimmer’s eyelid syndrome as well as histopathologic analysis from one patient. It appears that traumatic neuroma is the cause of the mass effect in swimmer’s eyelid.

Report of a Case. A 22-year-old man had a 5-year history of bilateral upper eyelid masses, which was worse on the right side. The patient had been in competitive swimming for 9 years and used hard plastic swim goggles. He denied any previous episodes of facial or ocular trauma. An ophthalmic examination of visual acuity, pupillary response, and extraocular movements, as well as slitlamp examination showed no abnormalities. An external examination revealed bilateral, sub-brow nodules, located in the area of the supraorbital notch, with the right nodule being larger than the left (Figure 1). No point tenderness was elicited. Excision of the right nodule was performed under local anesthesia, and there was no numbness or paresthesia in the distribution of the fifth cranial nerve, first division, either before or after surgical treatment. Histopathologic analysis revealed a haphazard prolifer-
tion, including all elements of nerve fascicles: axons, myelin, Schwann cells, and fibroblasts (Figure 2). After 12 months the eyelid is without clinical recurrence.

Comment. Amputation, or traumatic, neuroma of the eyelid has been described in one previous report. Neuroma developed in the patient after a blunt periorbital trauma with a rock, which resulted in swelling and ptosis of the affected eyelid. Traumatic neuromata from blunt trauma are far less common than from penetrating injury or surgery.

In our practice we have seen at least 3 cases of young competitive swimmers with upper eyelid swelling and a nodule localized to the medial sub-brow region. We believe this syndrome is caused by the use of nonpadded, hard plastic swim goggles (Figure 3). Our case illustrates at least one pathologic lesion associated with this clinical entity. This interesting finding warrants further investigation via microscopic analysis of all surgical specimens obtained from patients with competitive swimmer’s eyelid syndrome. Treatment for this entity may consist of switching to a more padded style of goggle, discontinuation of goggle use, or surgical excision of the affected tissue.

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A look at the past . . .

Snellen prescribes a stenopaic glass for his keratoconus patients. Since vision is of wider range when the horizontal slit is broad, and of greater acuteness when the slit is narrow, he has constructed a wedge-shaped slit broad in the periphery and ending in a sharp point in the centre.