This Cornea Grand Rounds comprises three complex cases. The appropriate management of these cases was aided by the use of advanced corneal imaging techniques. The first cases obviated the continued need for immunosuppression in differentiating a pigmented calcific plaque from scleromalacia by Ultrasound Biomicroscopy (UBM). Similarly, the second case had a clear demonstration by UBM of the anatomic cause for chronic, recurrent hemorrhage after cataract extraction. Lastly, confocal microscopy was able to document the presence of a cellular infiltrate and the lack of other foreign material in a patient after corneal ring segment implantation.

Introduction: Elmer Tu, MD (Attending)

The first case involved a 62-year-old Asian female referred to the IEEI for dry eye syndrome and scleromalacia. Pigmented lesions of the sclera were noted in both eyes, which were thought to be scleral thinning. She was diagnosed with scleromalacia, plateau iris, and dry eye syndrome (DES); peripheral iridotomy was performed. The patient was seen by a rheumatologist, who ruled out rheumatoid arthritis, a common comorbidity with scleromalacia. She was prescribed low dose methotrexate for the presumed scleromalacia even though she remained completely asymptomatic (no photophobia, redness, pain). In 2006, the patient was referred to the Glaucoma clinic at the IEEI for UBM to rule out an iris cyst but the image was insufficient to evaluate scleral thickness. In 2007, the patient was referred to the Uveitis clinic to determine whether the patient had scleromalacia or scleritis.

Medical history. The patient had a history of osteopenia, dyslipidemia, and GERD. She was on Fosamax,Prevacid, aspirin, fish oil, a multivitamin, and methotrexate. The patient’s mother had diabetes mellitus but no glaucoma. The patient reported no smoking or drinking and a review of systems revealed some shoulder and lower back pain.

Visual exam. The patient’s vision was 20/20 -1 in the right eye and 20/20-2 in the left. Pupils were round and reactive in both eyes with no afferent papillary defect noted. Extraocular movements were full, as were the confrontation visual fields. Her intraocular pressure was 15 in both eyes. Slit lamp exam showed pigmented lesions of the sclera near the muscle insertion sites, medial more than lateral in both eyes. The iris was positive for peripheral iridotomy and the lens shows 1+ nucleus sclerosis. Fundus exam was unremarkable.

Differential diagnosis. Potential differentials to consider include: scleromalacia (perforans) which is associated with rheumatoid arthritis and may cause progressive thinning if untreated; necrotizing scleritis is associated with previous episodes of severe pain and redness and is characterized by large areas of thinning, which may progress if untreated; Bisphosphonate (Fosamax) induced scleritis; conjunctival malignant melanoma which is progressive, mobile and may involve large feeder vessels; uveal prolapse which is typically associated with a history of trauma, surgery, or pain; ochronosis which is an autosomal recessive deficiency of homogentisic acid oxidase leading to deposition of HGA and is usually heavily pigmented; scleral pigmentation (nevus of ota); or senile scleral (Cogan) plaque.

Ultrasound biomicroscopy showed normal scleral thickness as well as white deposit at the medial and lateral rectus insertion sites with echoic shadowing. This is consistent with calcium deposition and the diagnosis of senile scleral plaque. This benign condition occurs most commonly in those above age 70 (about 25%) and does not require treatment. As in this case, it can be confused with scleromalacia, but also melanoma and scleritis. UBM is an excellent tool to identify them and obviate the need for expensive work up and potentially toxic therapy. Norm (1975) proposed that senile scleral plaque may be caused by mechanical stress deteriorating the sclera at muscle insertions.
Background on Uveitis-Glaucoma-Hyphema Syndrome (UGH)

Patients typically present with the triad uveitis, glaucoma and hyphema, which are considered to be secondary trauma to the angle structures, iris or the ciliary body and which leads to chronic inflammation, neovascularization, and recurrent hyphemas. These often lead to intractable secondary glaucoma due to chafing of the iris by the IOL or erosion of the haptic through the iris or ciliary body. This typically occurred in the early 1980s among patients with closed-loop and rigid ACIOLs, but has also been reported in patients with iris supported or loose PCIOLs, particularly in the sulcus. UGH has also been reported in patients with AcrySof single-piece IOLs who experienced postoperative displacement of one haptic into the sulcus. This is theorized to be due to the AcrySof design, specifically the flexible, thick haptics, squared optic/haptic edges and unpolished side walls.

Patients with UGH typically present with pain, photophobia, red eye, and decreased visual acuity. They may also demonstrate increased intraocular pressure, anterior chamber cell/flare, hyphema, and corneal edema. Potential other causes of spontaneous hyphema include: neovascular glaucoma, neovascularization of the iris, neovascularization of the angle iris lesions, or Swan syndrome.

Evaluation. A full ophthalmological history and exam with attention to the cornea, intraocular pressure, anterior chamber, iris and dilated exam, as well as the use of gonioscopy/UBM (to visualize the interaction of the IOL with the iris and ciliary body) may help in isolating the diagnosis. Management typically involves PredForté, Atropine, controlling the intraocular pressure as needed, but usually surgery is necessary to remove or exchange the IOL. The latter may prove difficult due to posterior synechiae or open posterior capsules.
Post-LASIK Ectasia: Sing Your Li, MD (Resident)

A 36-year-old Hispanic male was referred to the IEEI for evaluation of possible keratoconus. The patient had undergone bilateral LASIK in Mexico in 2003 and noted a bilateral decrease in vision (right more than left) after the surgery. His vision, however, remained stable until 2005, when he noted a gradual decline bilaterally. His medical and ocular history were otherwise unremarkable.

Exam. On exam, best corrected visual acuity was 20/30 in the right eye and 20/25 in the left. His cornea demonstrated inferior thinning and bowing in both eyes. The remainder of his anterior segment exam and funduscopic exam was non-contributory. Central corneal thickness was 442 in the right eye, and topography showed inferior steepening with a pattern of a claw-shaped deformity in the right eye. The left eye also demonstrated inferior steepening, despite being relatively asymptomatic.

Differential diagnosis. In general, areas of corneal steepening after LASIK may be the expected result of treatment to correct hyperopia, a central island of steepening in a myopic treatment zone, progression of pre-existing keratoconus following LASIK, or iatrogenic post-LASIK ectasia. Though claw shaped patterns are classically related to pellucid marginal degeneration (PMD), they may also be found in keratoconus and ectasia secondary to LASIK (Lee et al., 2007). In PMD, the area of maximal thinning is in the periphery inferiorly, with the area of corneal protrusion found above the area of thinning. This is distinct from keratoconus, in which area of protrusion is central or paracentral and coincides with the area of maximal thinning. In this patient, the most likely diagnoses were native keratoconus that progressed following LASIK, or corneal ectasia secondary to LASIK.

Management. Three months after the initial presentation at the IEEI, an intracorneal ring segment (Intacs) was surgically placed in the patient’s right cornea inferiorly via femtosecond laser (INTRA LASE). Post-operatively, he was started on moxifloxacin, prednisolone acetate, and Acular eye drops. The following day, the patient was doing well and his uncorrected visual acuity had improved one line in the right eye. The patient did not attend his next scheduled appointment, but was reportedly doing well and asymptomatic. At our insistence, the patient returned for a visit on post-operative day 11, reporting mild photophobia. He had run out of the prescribed moxifloxacin eye drops two days prior. At this point, his vision was stable but an infiltrate in the channel was noted in the right eye. This was presumed to be a bacterial channel infection and the corneal suture removed, cultured. The patient was started on frequent moxifloxacin and vancomycin eye drops (every hour). The bacterial keratitis improved on antibiotic drops and his vision remained stable. The cultures of the suture remained negative.
Background on Post-LASIK Ectasia

Post-LASIK ectasia results from structural weakening of the cornea either from the creation of a flap or from stromal ablation during LASIK. It is characterized by progressive steepening centrally or inferiorly and is associated with progressive thinning, unstable refractive error, and high astigmatism. Post LASIK ectasia can occur within weeks to 45-months after LASIK. Possible risk factors including thin stromal bed after stromal ablation, pre-op ectatic disease (e.g., subclinical keratoconus, pellucid marginal degeneration), high myopia, and thinner corneas.

In many cases, corneal ectasia secondary to LASIK can be difficult to distinguish from cases of native keratoconus. The clinical and topographic findings can be identical. Keratoconus is a progressive, non-inflammatory condition in which the cornea is also weakened and thins progressively centrally or paracentrally and assumes a conical shape. Treatment options for post-LASIK ectasia and keratoconus are similar. They include spectacles and rigid gas permeable contact lenses. If the patient is intolerant of contact lenses, surgical options include penetrating keratoplasty, intracorneal ring segments, or lamellar keratoplasty. Further stromal ablation is typically avoided.

Background on Intracorneal ring segments

Intracorneal ring segments were marketed initially as a means of correcting myopia by structurally reshaping the cornea. In 2001, Intacs were first used for the treatment of mild to moderate keratoconus (Colin et al., 2003), correcting the best corrected visual acuity to such an extent that the need for penetrating keratoplasty was delayed, if not eliminated. Intacs have also been used off-label in post-LASIK ectasia (Kymionis et al, 2003 and 2006) and have resulted in a stable improvement in best corrected visual acuity, spherical and cylindrical error, and corneal topography.

Potential adverse events associated with Intacs include anterior chamber perforation, microbial keratitis, implant extrusion, shallow ring placement, and corneal thinning. Microbial keratitis is found in less than 1% of patients, is associated with a broad range of pathogens, and can occur as late as 22 months after surgery, necessitating close follow-up (Hofling-Lima et al., 2004).

References


