Ocular Surface Disease and Irregular Cornea Grand Rounds: A Multidisciplinary Approach

Ocular surface disease with or without corneal irregularity is challenging to treat both optically and physiologically. This expert “all-Chicago” panel will present challenging cases and treatment approaches including medical, surgical, and contact lens management of corneal disease. The multidisciplinary approach to management of these patients will be evident as AAO Diplomates and Fellows present cases shared with internationally recognized corneal surgeons.

Each Case will present the underlying disease process, systemic manifestation in eye disease, medical, surgical and contact lens treatment approaches.

Objectives:
1. Learn of different ocular surface conditions that benefit from contact lens management
2. Understand the pathophysiology of various ocular surface diseases: Limbal stem cell disease, herpetic corneal disease, ocular manifestations of rheumatoid arthritis, and Stevens-Johnson Syndrome
3. Learn the medical and surgical management of the above conditions

Speakers:
Moderator: Loretta Szczotka-Flynn OD, PhD
Timothy McMahon OD
Charlotte Joslin OD, PhD
Ann Laurenzi Jones OD
Louise Sclafani OD
Marian Macsai MD
Dmitri Azar MD

1. Herpetic Corneal Disease
   a. Case Presentation
      i. neurotrophic non-healing persistent epithelial defect (no resolution multiple surgical options), resolved with scleral lens.
      ii. increased lipid deposits/denser central opacity with stromal vascularization and pt interested in PKP 2nd cosmesis – ultimately deferred due to poor outcome in neurotrophic eyes
      iii. VA/cosmesis improvement with increased steroids – pt remains stable and functional in scleral lens.
      iv. potential fitting issues contributing to increasing NV
   b. Review of pathophysiology and Treatment Approaches
      • Keratitis caused by HSV, most common cause of corneal blindness in developed nations.
      • Previously thought HSV-1 predilection for the trigeminal ganglion; HSV-2, for the sacral ganglion.
      • Increasing number of cases of ocular herpes are caused by HSV-2
      • Primary HSV infection occurs by direct contact with infected secretions. May be latent.
May replicate and travel back down the nerve to cause a primary infection that is clinically evident in 1% to 6% of infected patients.

Uncertain whether ocular recurrences are caused by virus that initially infected ocular tissues or by "back-door spread," via the trigeminal ganglion, from an initial oral infection.

Recurrent HSV infection most frequently involves the cornea, although all other parts of the eye can be affected concurrently or independently (retinitis, trabeculitis, uveitis, and optic neuritis)

HSV Epithelial and Stromal Keratitis

Epithelial Keratitis
  - Usually caused by actively replicating virus.
    - Dendritic ulcer
    - Dendritic scar, ghost dendrite, may remain in the superficial stroma.
    - Geographic ulcer; much larger epithelial defect.
      - In compromised immunity, especially patients taking topical corticosteroids.
    - Metaherpetic (trophic) ulcer is the only form of epithelial ulceration that does not have any live virus.
      - inability of the epithelium to heal

Stromal Keratitis
  - Immune-mediated response to nonreplicating viral particles,
  - Inflammatory response to viral antigen in the corneal stroma and can manifest as focal, multifocal, or diffuse stromal opacities.
  - If accompanied by corneal vascularization, interstitial keratitis
    - Novel treatment approaches
      1. anti-VEGF
    - long term prophylaxis

PKP outcomes in HSV
  - Compare and contrast success with PKP in neurotrophic corneas
    - experiences of Dr. Azar and Macsai

2. Limbal Stem Cell Deficiency (LSCD)
   a. Case Presentation
      - Diabetic neurotrophic ulcer, Limbal Stem Cell Deficiency, s/p Glaucoma Drainage Implant (posterior tubes) with non-healing persistent epithelial defect (PED) despite surgical options, resolved with scleral lens.
      - Consideration for LSC graft vs. PKP vs. combo after PED resolution – but eventually VA improved to functional vision and stable at present with scleral lenses.
   b. Review of Pathophysiology of LSCD
      - Causes in the absence of CL wear
      - Treatment options
        1. LSC graft
        2. PKP
        3. Combo of above
4. Can contact lens improve LSCD?

c. PKP outcomes in LSCD
   i. Compare and contrast success with PKP in LSCD with or without LSC graft
   ii. Dr. Azar and Macsai experiences

3. Stevens-Johnson Syndrome

   a. CASE REPORT INFORMATION
   
   • 19 yo WM 2006
   • Urinary tract Infection
   • Prescribed oral Bactrim for prostatitis
   • The initial 10 day course was not effective, given an additional 30 days
   • Rash on his neck started, then spread to his back. Several visits to the ED and steroid tx. did not resolve and he presents....
   • Toxic Epidermal Necrosis- Transfer to the Burn Unit

   b. PATHOPHYSIOLOGY OF STEVENS-JOHNSON SYNDROME
   
   • Rare, serious disorder of skin and mucous membranes.
   • Initial flu-like symptoms, tongue swelling, hives, then painful red or purplish rash that spreads and blisters.
   • Top layer of the affected skin dies and sheds.
   • Admitted to Burn Unit for palliative care, supportive therapy and infection control
   • RISKS:
     • Immuno-suppression, past or family history, HLA-B 1502 gene
   • Reaction to a medication
     • Anti-gout medications
     • Pain Medications
     • Antibiotics
     • Anticonvulsants /antipsychotics
     • Radiation therapy
   • Reaction to Infection
     • Herpes
     • Pneumonia
     • HIV
     • Hepatitis

SJS COMPLICATION

• Secondary skin infection (cellulitis)
• Blood infection (sepsis)
• Internal organ damage
• Permanent skin damage
• Eye problems:
  • Inflammation
  • Irritation/dry eyes
  • Eyelid inflammation
• Scarring
• Blindness

c. TREATMENT OPTIONS FOR OCULAR INVOLVEMENT

TARSORRHAPHY
Surgically close the palpebral fissure by suturing the superior and inferior lids at the lateral aspect

STAMLER LID SPLINT
Adhesive on one side with enough rigidity on the other to hold the eyelid in the closed position

TAPESORRHAPHY
Tagederm
Allow for use of meds and examination

AMNIOTIC MEMBRANE
For severe cases such as Stevens Johnson Syndrome and chemical burns
The Classic Ring – acts as a symblepheron ring to maintain the orbital space
Multiple layers of tissue – results in additional therapeutic function by staying on the eye longer to maintain biologic action and durability to reduce inflammation and promote healing and longer biologic action on the ocular surface

BANDAGE CONTACT LENS
• Confirm Diagnosis
• CONSENT/RESPONSIBILITY
• Careful/tailored follow-up
• Minimal movement but monitor for edema
• AB prophylaxis for acute ddx
• Non-Preserved Lubricants
• Sterile technique
• DO NOT REMOVE TOO EARLY
• LONGTERM SAFETY STUDIES
  • Wills N= 74, retro, 2006-09
  • Oasys or Night/Day
  • PBK, PK, EBMD, LSCD, DES, NT, BK, SLK
  • Pro AB= 73%-88% FQ tid
  • WT = 21 – 235 days
  • MK: 2 %, 2 cases
  • +AB, Steroid, epi-defect
  • RCE after 272 days (r=6)
  • LSCD after 2 days

MORE ON THIS PATIENT CASE
• SJS 2006 after Bactrim
• Bandage Contact Lens RXED (2006)
• Lost to Follow-up: weekly EW… becomes PRN
• Neovascularization/ GPC develops (Q4 2012)
• Microbial Keratitis- fungal (Q1 2013)
• Corneal Scarring
• Refit in to Scleral GP: Notching due to pyogenic granuloma

PATIENT COMPLICATION: PYOGENIC GRANULOMA

• AKA lobular capillary hemangioma
• Small, round, hyperplastic vascular lesions
• Not Pyogenic or granulomatous, benign
• Result of trauma, inflammation, bug bite, hormone (pregnancy)
• Predisposing factors: corneal ulceration, dry eye syndrome, alkali burn, trachoma, trichiasis, post radiation
• Rapid growth, stabilization or regression, recurrence

• Continuous Wear of above 2 yrs. “successful”
• Outside provider re-fit into BCL and develops MK in 2 weeks
• MEETS WITH CORNEA SPECIALIST who requests:

  Elevation Specific Technology
  • Definition and Development
  • Indications and Applications
  • How to take an Impression: Do’s/Don'ts
  • Design the Prosthetics
  • Trouble-shooting

• Compare and contrast other treatment options with Dr. Macsai and Azar

4. Rheumatoid arthritis with ocular complications: ulcer formation after Cataract Extraction

  a. Case Presentation
     1. Plugs and Restasis
     2. Relief of Dry eye and vision improvement with scleral lens
     3. Difficult insertion and removal from RA affecting hands
     4. VIDEO

  b. Review of Complications and Pathophysiology of Rheumatoid arthritis (RA) and treatment of RA
     i. RA with ocular manifestations
        ▪ Peripheral ulcerative keratitis PUK, corneal melt from immune complexes infiltrating vasculcar arcade
        ▪ RA most common etiology (34% of cases)
        ▪ Stromal thinning and overlying epithelial defect
        ▪ Neovascularization
        ▪ Can have pain, photophobia, tearing, irregular astigmatism
        ▪ Increased mortality in RA patients, can be sign of impending vasculitis
     ii. Associated surgical considerations for CE and PK in a pt with RA.
     iii. Treatment
- Management of underlying RA
- Systemic corticosteroids with or without immunologic agents
- Ocular, treat coexisting dry eye
- Tissue adhesives
- Amniotic membrane
- Bandage contact lens
- Tectonic, lamellar transplants
- Scleral lens

c. Compare and contrast treatment options with Dr. Macsai and Azar