Carrasquillo Outline

When lightning strikes twice: Seven Cases of Stevens-Johnson Syndrome and Corneal Ectasia

Abstract
Corneal vascularization, scarring, thinning, trichiasis and conjunctivalization are often ocular complications of Stevens Johnson syndrome. In cases of SJS and corneal ectasia, ocular management proves to be challenging. We review the course and management of seven cases with concomitant ocular surface disease and corneal ectasia.

History
- Retrospective medical record review of cases extracted by search on diagnoses of SJS and corneal ectasia on manufacturing database of PROSE devices fabricated from 2002-2011.
- Review of history on causal vs. coincidental relationship between these syndromes was conducted.
- All patients were referred to the Boston Foundation for Sight for treatment of their ocular surface disease and for their visual rehabilitation.
- All patients had attempted soft lenses, gas permeable lenses, piggy-back lenses, and/or hybrid contact lenses without success; poor tolerance.

Pertinent Findings
- Seven patients 5F:2M
- All with SJS/Corneal Ectasia diagnosis
- Average age at SJS diagnosis – 15 years old.
- Average age at Ectasia diagnosis – 22 years old.
- Most patients were diagnosed with corneal ectasia post SJS.
- All patients received PROSE treatment for reduced vision and visual function (13 eyes of 7 patients)
- Best corrected visual acuity on presentation ranged from 20/25 to counting fingers (CF) with various modes of correction, including specialty contact lens.
- Visual acuity improved in all eyes. All patients achieved driving vision (≥20/40)
- Average wear time of PROSE device ranged from 8-16 hours.
- Improvement in visual function was demonstrated both subjectively and as measured by NEI VFQ-25
- All patients were fitted successfully with PROSE devices

Diagnosis and Discussion
- Stevens-Johnson Syndrome can lead to severe ocular complications during chronic stage of disease:
  - Scarring of conjunctiva
  - Symblepharon formation
• Entropion
• Trichiasis
• Tear film disturbances
• Corneal involvement
  i. Scarring
  ii. Neovascularization
  iii. Keratinization
  iv. Thinning
  v. Conjunctivilization
• Lacrimal Duct involvement
  i. Cicatrization
  ii. Destruction of Goblet Cells
• Limbal stem cell deficiency
  o SJS eyes often develop:
    • Corneal Opacities
    • Corneal erosions
    • Epithelial breakdowns
    • Ulceration
    • Infectious Keratitis
  o Because of limbal stem cell deficiency, SJS eyes are poor candidates for corneal transplants.
  o Corneal ectasia:
    • Decreased best corrected visual acuity
    • Often times spectacle correction or soft lens correction are not viable options
    • Require specialty contact lenses; rigid gas permeable (GP) lens correction
    • For highly ectatic and irregular corneas, corneal GP lenses may be unstable
    • Often if complications with GP lenses, alternative approach is corneal transplants.
  o Concomitant SJS and corneal ectasia is particularly challenging:
    • Traditional GP lenses for ectasia visual rehabilitation are poorly tolerated in SJS patients
    • Option of penetrating keratoplasty is not viable one in concomitant cases of SJS

**Treatment and Management**
  o Prosthetic replacement of the ocular surface system (PROSE) treatment
    • Use of scleral prosthetic devices.
  o These are FDA-approved custom designed and fabricated prosthetic devices:
    • Restore vision,
    • Support healing
    • Reduce symptoms
    • Improve quality of life for patients suffering with complex corneal disease
• Large diameter (average 15.5–23 mm)
  ▪ Designed to rest entirely on the sclera
  ▪ Vault entirely the cornea
  ▪ Creating a space filled with preservative-free artificial tears or saline
  ▪ They are fluid-ventilated
  ▪ Allow the containment of an oxygenated pool of artificial tears free of air bubbles over the cornea

  o This case:
  ▪ All patients were custom-fitted with a scleral prosthetic device
  ▪ Devices were worn on a daily wear schedule
  ▪ Improved best corrected visual acuity and visual function in all eyes.
  ▪ Significant decrease in dry eye symptoms, pain, and photophobia.
  ▪ In five out of seven patients, ectasia developed after SJS onset.
  ▪ PROSE treatment:
    ▪ Adequate ocular surface support in refractory disease
    ▪ Viable and excellent choice for both SJS and corneal ectasia
      i. Excellent treatment option for concomitant cases of both complex ocular surface disease and ectasia
    ▪ Passive and non-invasive treatment
      ii. No surgical intervention
      iii. Potentially precludes need for further surgical intervention

Conclusion
  o Patients with corneal ectasia and concomitant ocular surface disease from Stevens-Johnson syndrome pose a special challenge for visual rehabilitation.
  o Contact lens may be poorly tolerated, difficult to fit, and penetrating keratoplasty is high risk.
  o PROSE treatment is a useful option for patients such as these with complex corneal disease.
  o There is not enough data in the literature to confirm a causal vs. coincidental relationship between SJS and corneal ectasia.
  o The fact that 5 out of the 7 patients in our series were diagnosed with ectasia some time after SJS diagnosis may suggest that severe ocular surface microtrauma maybe a factor in developing ectasia.
  o More data is needed to further conclude on a causal vs. coincidental relationship between SJS and corneal ectasia.

References
1. Rosenthal P, Croteau A. Fluid-ventilated, gas permeable Scleral contact lens is an effective option for managing severe ocular surface disease and many corneal disorders that would otherwise require penetrating keratoplasty. Eye & Contact Lens. 2005; 31(3) 130-134.


Abstract:
Despite rare indications for prosthetic iris implantations, there are complications that need prompt recognition and treatment. This case depicts complications of the procedure, devastating course of disease, and importance of patient education on proper management and treatment.

I. Case History
A. 32 YO white female presents for first time at VA eye clinic in Chicago
   1. Chief complaint is dissatisfaction of color of cosmetic iris implants, dryness, and glare at night.

B. Past ocular history:
   1. Bilateral PRK in 2005
   2. Bilateral dry eyes
   3. Bilateral iris implants

C. Past medical history/social history:
   1. Anxiety
   2. Hyperlipidemia
   3. Liposuction in face
   4. Breast implants
   5. Rhinoplasty
   6. Eyebrow lift
   7. Lip surgery

D. Medications:
   1. Simvastatin
   2. Clonazepam

E. Other Salient Information
   1. Allergy to codeine with reaction of emesis

II. Pertinent Findings
A. Clinical
   1. Visual Acuities (without correction)
      a. OD: 20/20
      b. OS: 20/20

   2. Direct pupillary response OD, however, pupil reaction OS could not be determined due to blockage of pupillary frill from implant. Consensual pupillary response and presence or absence of afferent pupillary defect could not be assessed.

   3. EOMs: NL, CVF: Full to Finger Count OD/ OS
   4. Cornea: tr SPK OU, 3+ dispersed endothelial pigment and epithelial scars OU. Anterior chamber: occasional pigment cell OU. The patient’s true iris was
minimally visible behind the green intraocular implant of artificial iris OD, but not visible at all OS
5. IOP: OD 28mmHg, OS 28mmHg at 9:05am by applanation
6. Lens: clear to extent seen OU
7. Dilated Fundus Exam:
   a. OD: CD: 0.3
   b. OS: CD: 0.3
9. Gonioscopy: Ciliary body in all quadrants with significant pigment in the trabecular meshwork with areas of implant obstructing views OU
10. HVF: OD: full, OS: full

B. Physical
1. BP: 120/78 right arm sitting.
2. Pulse 69 bpm; strong, regular
C. Lab work including CBC, thyroid, and lipids: NL on medication

III. Differential Diagnosis
A. Primary/Leading – Ocular hypertension OU secondary to cosmetic eye surgery

IV. Diagnosis and Discussion
A. Dry eyes OU s/p PRK OU
B. Ocular Hypertension s/p intraocular implant of artificial iris OU. Thin CCT, but NL VF OU.
C. Anterior nongranulomatous iritis OU s/p intraocular implant of artificial iris
D. The reason for adverse effects in implants anterior to the iris is likely directly related to chronic inflammation, pigment dispersion, and compression of the trabecular meshwork by the implant and indirectly to chronic steroid use.
E. Indications for iris implants, despite there being no FDA approval, include trauma, post-excision of iris melanoma, iris coloboma, congenital aniridia, iridocorneal-endothelial syndrome and other inflammatory causes of iris damage
F. Risks included intraoperative development of hyphema, suprachoroidal hemorrhage and retinal detachment, and post-operative cataract development, IOP increase, corneal edema/decompensation, uveitis, or endophthalmitis.
G. Explantation should be performed at the most earliest sign of any complication

V. Treatment Management
A. AT QID OU
B. Begin Travatan qbedtime OU
C. Begin Brimonidine bid OU
D. Pt educated to have cosmetic iris implants explantation
E. Bibliography

20. C.Y.W. Khng and M.E. Snyder., “LE216 functional and cosmetic outcomes after artificial, iris implantation” The Eye Institute, Tan Tock Seng Hospital, Singapore. Cincinnati Eye Institute, Cincinnati, Ohio, USA
22. Arundhati, Anshu, et. al, “Iris Reconstruction in Penetrating Keratoplasty—Surgical Techniques and a Case-control Study to Evaluate Effect on Graft Survival”, Singapore National Eye Centre, Yong Loo Lin School of Medicine, National University of Singapore, Singapore, Singapore Eye Research Institute, Yong Loo Lin School of Medicine, National University of Singapore, Singapore, Department of Ophthalmology, Yong Loo Lin School of Medicine, National University of Singapore, Singapore, Accepted 2 October 2007. Available online 25 January 2008.

VI. Conclusion
A. Although iris prosthetic devices do provide benefits, they can and do have destructive results. Close and regular follow up is important to avoid such complications. As with any medical procedure, risks and benefits should be properly assessed. Depending on the reason for treatment, iris implant risks may outweigh benefits, especially in cases for cosmetic purposes given the serious complications of the implant and removal of implant. However, in contrast, a patient with severe symptoms secondary to congenital or acquired defects the benefits may outweigh the risks. This case portrays a young patient who had healthy eyes that rapidly changed to diseased eyes as result of opting for cosmetic iris implants. Patient education continues to be essential on proper treatment and management.
Choroidal Retinal Lesion in a Patient with History of Prostate Cancer (A New Meaning to Flashes) Annie Wan

**Case Report**

A 73 year old Caucasian male presents for a routine glaucoma examination, but reports symptoms of a persistent flash in his left eye. Clinical exam reveals a choroidal lesion suspicious for melanoma or metastasis.

**Case History**

8/2010 70yo WM presents in August 2010 for initial examination with no ocular or visual complaints. His last reported dilation was more than 40 years ago at which time he was told there was a spot in the back of his eye. He denies any family history of ocular conditions, history of trauma, or ocular surgery. Hypertension since 2002: Hydrochlorothiazide, Lisinopril (132/70, rate 76)

Diabetes Mellitus: Metoprolol

Hypercholesterolemia: Simvastatin

Low compound hyperopic prescription with best corrected vision of 20/20 OD, OS.

Pupils, ocular motilities, confrontation fields, and Amsler grid tests were unremarkable.

Anterior segment evaluation showed slight capped meibomian glands and mild cataracts OU.

Intraocular pressures were measured to be 16mmHg OD and 17mmHg OS.

Dilated exam revealed moderate cupping of the discs 0.65 OD and 0.60 OS, moderately attenuated blood vessels with few crossing changes, and a 3.5 disc diameter flat choroidal nevus located superonasal OD. All other findings were unremarkable.

Patient was diagnosed with blepharitis, mild hypertensive retinopathy, moderate optic nerve cupping, early cataracts, and choroidal nevus OD. The patient to return in 3 months for baseline visual fields.

9/2011 71yo: CC itchy/tearing eyes and symptomatic relief with Visine

Recently diagnosed and undergoing radiation treatment for prostate cancer since July 2011.

Pain: Acetaminophen, Ibuprofen

Gout: Allopurinol

HTN: Amlodipine, Hydrochlorothiazide, Lisinopril (116/78, rate 64)

GERD: Omeprazole

CHL: Simvastatin

Prostate: Terazosin, Warfarin

Inactive: Bicalutamide, Colon elec lavage, fentanyl citrate injection, midazolam IV injection

Intradermal injection: Enoxaparin, Goserelin

Intraocular pressures were 19/18 with cupping being noted as 0.70 OD, 0.75 OS.

Choroidal nevus OD was stable.

Return in 3 months for visual fields, glaucoma work up.

12/2011 72yo WM Humphrey visual field 12/2011 was reliable and full OD, reliable with superior arcuate defect OS not noted from 2010. Gonioscopy revealed open angles. Corneal thickness was average (544/540). Diagnosed with primary open angle glaucoma, started with travatan qPM OU, and return in 3 months for repeated fields. Last measured IOP 14/14. Choroidal Retinal Lesion in a Patient with History of Prostate Cancer (A New Meaning to Flashes) Annie Wan AAO 2013
8/2012 72yo WM, Ran out of travatan x 3 weeks. Last visual fields (5/2012) was stable OU.

Pain: Acetaminophen, Ibuprofen

Gout: Allopurinol

HTN: Amlodipine, Hydrochlorothiazide, Lisinopril, metoprolol (130/78, rate 66)

GERD: Omeprazole

CHL: Simvastatin

Prostate: Terazosin, Warfarin, Tamsulosin

Inactive: Bicalutamide, Colon elec lavage, fentanyl citrate injection, midazolam IV injection

Intradermal injection: Enoxaparin, Goserelin

IOP 15/16

Choroidal nevus OD is slightly elevated on clinical observation.

Return in 3 months for pressures.

12/2012 73yo WM, POAG returning for pressure check, travatan qPM OU with good compliance

Reports “light bulb” inferiorly OS for 1 month. Denies floaters/curtains.

IOP 18/19, C/D 0.70 thin sup OD, 0.75 thin inf OS

OD: Choroidal nevus flat sup

OS: 4DD choroidal lesion (+)drusen, elevated, pigmentary changes

B-scan shows small elevation with minimal internal reflectivity

FA confirms early hyperfluorescense that does not change in the late stages. (+)allergy to fluorescein

Referral to outside ocular oncologist immediately.

IMAGES 2012: OD NEVUS OS CHOROIDAL LESION

Differential Diagnosis

- Elaborate on condition
  - Choroidal melanoma is most common primary intraocular malignant neoplasm in adults
    - 88% choroidal, 12% ciliary body/iris
  - Cancer that arises from melanocytes
    - Melanoma 4% skin cancers, 79% skin cancer related deaths

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Secondary cancers: 5yr ~7.7% rate, most common prostate (23%), breast (17%)  
6% prostate cancers metastasize to the choroid  
50% mortality rate  
- 33% from melanoma metastasis, 29% malignant tumor other than metastatic, 11% malignancy of unknown origin  
- Median survival for stage 4 ~9mos, and 3yr survival <15%  
- Poor prognosis 70-79yo, males>women 3:2 >80yo  
Diagnosis/Staging scales  
- LUMPO  
- TNM – AJCC and UICC  
- Kaplan-Meier analysis  
- Early diagnosis = surgical resection (80% effective for thin lesions)  
- Other presentations: Subfoveal melanoma → Tx plaque brachytherapy → 
- Expound on unique features  
- Risk of metastasis depends on several factors: gene expression, basal tumor diameter, tumor thickness, ciliary body/extraocular muscle involvement, melanoma cytomorphology, etc.  
- Diagnostic tests  
- Transcleral fine needle aspiration biopsy  
- Cell culture: cell lines, blood DNA  
- B scan Ultrasonography  
- Fluorescein Angiography  
- Indocyanine Green Angiography  

Treatment/Management  
- Treatment  
- Enucleation  
- External proton beam radiation  
- Transcleral local resection  
- Endoresection  
- Transpupillary thermotherapy  
- Plaque radiation therapy  
- Iodine Brachytherapy  
- Androgen deprivation therapy: Gonadotropin-releasing hormone agonist (Leuprolide acetate)  
- Intralesional injection BCG/interferon/interleukin 2 (stage 3)  
- Dacarbazine, Temozolomide, high dose IL2, and paclitaxel (stage 4)  
- Alternative Photodynamic therapy (PDT): induce direct tumor cell photodamage, destruction of tumor vasculature and activation of an immune response  
- Photosensitizing agent + illumination of the tumor with visible light  
- Apoptosis = controlled, energy consuming process of suicidal cell death  
  - Mitochondria mediated/intrinsic pathway  
  - Death receptor-mediated/extrinsic pathway  
- Necrosis
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- Protective mechanisms against therapy: pigmentation and increased oxidative stress defense
  - Response to treatment
    - Visual symptoms improve significantly depending on location

**Conclusion**
- Clinical Pearls
  - Lesions present when you least expect them
  - Prompt referral important
  - Ancillary tests like OCT serves a valuable tool
  - Discussion about prognosis important
  - Most aggressive treatment may not be the best
  - Other malignancies may occur

**Bibliography**