Management of Severe Ocular Surface Disease Secondary to Stevens-Johnson Syndrome with BostonSight® PROSE Treatment

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Abstract:
A thirteen year old Hispanic female presents with severe ocular surface disease secondary to Stevens-Johnson syndrome (SJS). Successful BostonSight® PROSE treatment improves visual acuity and photophobia Utilizing moisture goggles filled with saline, eased device adaptation.. (35 words)

I. Case History
- CM a 13 year old Hispanic female
- Chief complaint: severe dry eye OU with constant burning, redness, pain and light sensitivity over the last 6 years. Symptoms are worse in the morning and improve mildly with eye drops
- Pt has a history of poor academic performance and difficulty with reading for extended periods.
- History of Stevens- Johnson syndrome (SJS) in 2009 due to Amoxicillin
- Ocular History: corneal and conjunctival scarring OU, entropion RUL and LUL, keratoconjunctivitis sicca OU and symblepharon OU
- Medical History: Asthma, SJS 2009
- Medications: Symbicort 160/4.5 mcg aerosol inhaler 2 puffs PO BID
  Ventolin HFA 90 mcg aerosol inhaler 2 puffs PO QID
- Eye drops: Genteal gel 0.25% 1 gtt QHS OU, PFATS 1 gtt QID OU
- Allergies: Amoxicillin

II. Pertinent findings

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<td>OD: NI</td>
<td>OS: 20/200</td>
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<td>OD: &lt;20/800</td>
<td>OS: 20/400 @6in</td>
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<td>CC PROSE Near</td>
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- Slit lamp findings:
  8/3/15 Initial Exam
- Lids/Lashes: Entropion UL, keratinized lid margins OU, trichiasis OU
- Conjunctiva: OD: 2+ injection, sup symblepharon up to sup limbus
  OS: 2+ injection, sup symblepharon
- Cornea: OD: dense central stromal scarring, neovascularization 360 w/ central haze and semi-opaque cornea, diffuse 3+ SPK, no epi defects or infiltrates
  OS: dense superior stromal scar, neovascularization entering visual axis,
diffuse 3+ SPK, no epi defects or infiltrates
- Iris: OD: poor view; OS: flat, clear
- Anterior chamber: OD: poor view, OS: Deep and Quiet
- Lens: poor view OU

Final PROSE Device OS: Vault: 4.6mm, BC: 7.9mm, Power: +2.50 D, 16.5mm diameter, Boston XO2

III. Differential diagnosis

Diagnoses:
- Steven's Johnson syndrome w/ significant ocular involvement
- Dry eye syndrome OU- severe (OD>OS)
- Corneal Opacity OD>OS
- Corneal Neovascularization OU

Differential Treatment options:
- Continue with current treatment
- Punctal plugs or cautery
- Restasis
- Non-preserved artificial tears
- Autologous tears
- Amniotic membrane
- Surgical tarsorrhaphy

The current treatment methods have failed to address the severe dry eye symptoms. PROSE treatment was selected for the benefits of allowing the surface to be protected from further insult while also improving visual function and comfort.

IV. Diagnosis and discussion

- Stevens-Johnson Syndrome: a rare immune complex modulated hypersensitivity reaction that attacks the skin and mucous membranes. This condition is potentially life threatening.
- SJS can be induced by medications, infectious agents or malignancies
- Drug-induced cases are most commonly linked to antibiotics followed by anti-seizure drugs and non-steroidal anti-inflammatory drugs (NSAIDs) (1).
- Initial presentation can resemble an upper respiratory infection featuring fever, erythematous burning rash, fatigue and productive cough.
- Necrotic skin lesions cause separation of the epidermis from the dermis and results in sloughing of skin.

Classification is based on body surface area involvement:
- Erythema minor: skin involvement only
- Stevens-Johnson Syndrome (SJS): <10% of body surface area (BSA)
- SJS/TEN overlap: between 10 and 30% BSA involvement.
- Toxic epidermal necrosis (TEN): >30% BSA involvement
- The level of skin involvement is proportional to mortality rate with this condition. Complications such as sepsis and bacteremia are life threatening.
- In acute phases, patients are managed similar to burn patients by treating skin lesions, managing pain and monitoring for infection.
- According to one study, only 11% of patients had ocular manifestations initially however, up to 89% of patients had chronic ocular complications (2).
- Late ocular complications include: (2).
  - **Cornea**: opacification, conjunctivalization, neovascularization, superficial punctate keratitis (SPK), and loss of palisades of Vogt (POS)
  - **Conjunctiva**: keratinization, scarring, symblepheron formation
  - **Eyelid**: entropion, keratinization, trichiasis

- The largest obstacle in this case was insertion and removal of the devices.
- Using moisture goggles filled with saline prior to insertion training helped to desensitize corneas to the saline. Prior to using the goggles the patient would frequently recoil as soon as the saline within the device would contact her eyes. We filled a pair of swimming goggles with non-preserved saline for 10-15min prior to insertion and removal training. This helped with the sensitivity of her lids and corneas.
- This particular case required roughly ten separate training sessions over the two weeks that PROSE treatment was being initiated.
- The other challenge was the significant symblepheron formation, especially in the right eye. This decentered the lens downward, even with smaller diameter designs and made it very difficult to assess the fit as well.
- There was no visual improvement with the device over the right eye and the fit was unsuccessful.
- Even with only the left device in place, there was an obvious change in the patient's posture in terms of being able to keep her eyes open comfortably for more time.

V. **Treatment, management**
- Successful insertion of PROSE device by mother's fiancé, patient able to remove device.
- Unable to fit PROSE device OD due to severe symblepheron and no improvements in acuity with treatment OD.
- Continue with PROSE treatment OS for daytime wear.
- Initiated FML ung QHS OU. Patient has historically had poor compliance with Pred Forte drops as prescribed by corneal specialist.
- Continue with PFATs QID OU.
- Consider fornix reconstruction with mucous membrane grafts of the lid margins to decrease constant irritation of the cornea.
- RTC in 2 months for progress evaluation.
- Significant improvements in visual acuity in the left eye: from entering 20/400 to 20/70-2 OS.
- PROSE also decreased photophobia and increased overall comfort in the left eye.

Treatment of ocular complications of SJS:
**Acute Phase treatment:**
- amniotic membrane transplantation has been successful in preventing the development of chronic ocular complications if initiated in the acute phase of this condition (3)
**Chronic treatment options (4).**
- Aggressive dry eye therapy i.e. Artificial tears, gels, ointments, cyclosporine 0.05%, punctal occlusion/cautery, moisture chambers, tarsorrhaphy
- Trichiasis management: epilation, electrolysis
- Entropion repair
- Mucous membrane grafting
- Conjunctival replacement surgery (COMET)
- Limbal stem cell transplantation
- Scleral Lenses or PROSE treatment
- Corneal transplant or Keratoprosthesis: poor prognosis due to underlying limbal stem cell deficiency, active inflammation and chronic dry eye.

Benefits of PROSE for SJS patients.
- Reduce dry eye symptoms of burning, tearing, pain and light sensitivity.
- Can improve visual acuity and function but masking irregularities of the cornea
- Has the ability to maintain the integrity of the ocular surface by protecting from further insult by the eyelids and lashes (5).

VI. Conclusion
- SJS is a rare condition that can cause drastic changes to the ocular anatomy and can lead to chronic ocular surface disease.
- This case is a good example of improvements that can be made in vision, function, comfort and maintaining the ocular surface using the BostonSight PROSE device.
- During the adaptation process, we utilized moisture goggles filled with saline to help the patient become comfortable with the saline in the devices. Insertion and removal of the devices is a challenge that was overcome with many training sessions.


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