Ocular outcomes in Toxic Epidermal Necrolysis Syndrome: a 13-year review at Chelsea and Westminster Burns Intensive Unit

Authors: M.SOBTI[1], O. Li[1], A. PANDEY[2], R. SCAWN[1], D. COLLINS[1], C. BAILEY[1], J.LEON-VILLAPALOS[1]

1. Chelsea and Westminster Hospital NHS foundation Trust
2. Imperial College Healthcare NHS Foundation Trust

INTRODUCTION

Steven Johnson Syndrome (SJS) and its more severe form, Toxic Epidermal Necrolysis (TENS) are rare, drug-induced, immune-mediated acute blistering diseases involving the skin and mucosal surfaces (UK incidence 1 per million/year).

Over half of patients exhibit ocular involvement, leading to severe cicatrising consequences and visual impairment (Figures 1-4).

Epithelial defects at the lid margin, conjunctiva and cornea are used to grade ocular involvement in TENS into mild, moderate or severe in order to guide treatment.

Intensive topical treatment with preservative-free steroids, lubricants and antibiotics are required in the acute phase. Amniotic membrane transplants (AMT) are recommended for moderate to severe cases. Systemic immunosuppression, although controversial, has shown benefit.

AIM

This is a retrospective case series of all patients treated for TENS at a tertiary burns unit in Chelsea and Westminster Hospital, London over 13 years.

We present our multidisciplinary experience of this rare illness.

METHOD

The records of patients treated for TENS at Chelsea and Westminster Hospital between 2010-2023 were accessed on Cerner software.

Data was collected on:

- Ocular involvement and its severity
- Dermatological status
- Patient demographics
- Causative agent
- Frequency of ophthalmology input
- Management of ocular involvement
- Ocular outcomes

RESULTS

- 24 patients developed TENS over 13 years

Ocular Involvement
- 66% (n=16) had ocular involvement
- 13% (n=3) were documented to have no ocular features
- 21% (n=5) had no data was available

Causative agent
- Lamotrigine (25%, n=6)
- Antibiotics (25%, n=6)
- Diazepam (4%, n=1)
- NSAIDS (8%, n=2)
- No identified trigger (4%, n=1)

Severity
- 44% (n=7) developed mild-moderate ocular disease acutely
- 56% (n=9) developed severe ocular disease acutely

Dermatological status
- Severe ocular disease correlated with higher TBSA (68% vs 81%) involvement
- Skin re-epithelisation took longer in severe ocular disease (7.2 vs 9.6 days)
- No differences were observed in SCORTEN between mild-moderate and severe disease groups

CLINICAL FEATURES

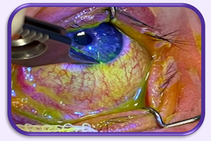


Figure 1. Extensive conjunctival de-epithelisation

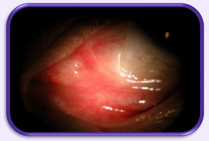


Figure 2. Symblepharon

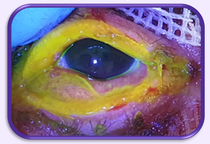


Figure 3. Severe lid margin de-epithelisation

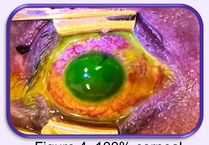


Figure 4. 100% corneal epithelial loss

TREATMENT RESPONSE

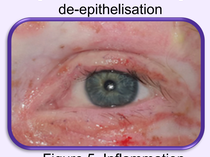


Figure 5. Inflammation settled with AMT

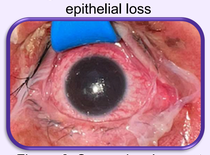


Figure 6. Severe involvement needing repeated AMT

SEQUELAE

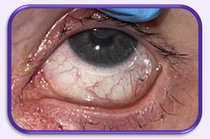


Figure 7. Corneal scarring



Figure 8. Severe Limbal Stem Cell Deficiency

RESULTS

Treatment
- 44% (n=7) were seen by an ophthalmologist within 24 hours
- 38% (n=6) were seen between 2-4 days
- 19% (n=3) had insufficient data

- All patients received systemic immunosuppression alongside steroid, lubricant and antibiotic drops.
- The severe ocular disease cohort all had cryopreserved AMT alongside medical management
- Additional topical agents were used on a case-by-case basis

AMT outcomes
- 25% (n=2) had resolution of inflammation and corneal epithelial defects (Figure 5)
- 56% (n=5) had sight threatening complications (Figure 6)
- 1 patient died
- 1 patient refused treatment

Sequelae
- 31% (n=5) had visual impairment (Figure 7 & 8)
- 31% (n=5) had severe chronic dry eye
- 31% (n=5) had no documented sequelae
- 1 patient died within 15 days of rash onset

CONCLUSIONS

In our study, nearly two-thirds of patients diagnosed with TENS exhibited ocular involvement, with two-thirds of this group experiencing long-term consequences. This data provides valuable insight for counselling patients on disease severity and potential outcomes.

Severe ocular disease presents considerable challenges and may lead to cicatrizing consequences despite AMT and receiving both topical and systemic immunosuppressive therapies. Multidisciplinary care is important for improving outcomes of this rare disease.

Future studies should be planned on:
- Optimal timing of AMT
- Correlation of dermatological and ocular manifestations
- Long term follow-up data of ocular outcomes

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