Abstract: Steven Johnson Syndrome SJS is an important cause of ocular morbidity. We present a series of 9 SJS patients who presented to our institution in the acute phase. All patients with the diagnosis of SJS over last 1 year were included. Possible etiologic factors, various ocular manifestations in acute phase is discussed. Early aggressive management included separating the lids frequently, cleaning the discharge, glass rod sweeping along the fornices and voluntarily moving the eyes. This prevented formation of symblepharon, ankyloblepharon and other sequelae. All patients recovered without any sight threatening sequelae. Mild sequelae included – one patient had ankyloblepharon and punctual stenosis, another had inferior corneal opacity, mild dry eye was seen in 9 cases. This study lays emphasis on the importance of early aggressive management in preventing and reducing the severity of sight threatening sequelae of SJS.

Keyword: Steven Johnson syndrome, symblepharon, ankyloblepharon.

Steven Johnson Syndrome [SJS] is a symptom complex characterized by symmetrically distributed erythematosus bullous lesions of the skin and mucous membranes. It is accompanied by severe constitutional symptoms and also called erythema multiforme major. It predominantly involves the oral mucosa and conjunctiva [ocular involvement 69 - 91% in adults]. It is essentially drug induced². Anticonvulsants, antibacterial, some non-steroidal anti-inflammatory drugs have been identified as probable causative factors. Other factors like viral infection, mycoplasma have been suggested to precipitate SJS. Ocular manifestation can be classified as mild with lid oedema, conjunctivitis, chemosis; moderate with conjunctival membranes, corneal epithelial loss and corneal ulceration; and severe with cicatrical changes and perforation. Prompt diagnosis, identification and early withdrawal of all suspect drugs are the most important² . The management of the patients must be undertaken in the specialized intensive care units. Ocular sequelae can be minimized by prompt, early and regular ophthalmic care. We present a series of 9 patients (Aged 20-50 years) who presented to our institution in the last 1 year.

Table 1. History of drug intake

<table>
<thead>
<tr>
<th>Name of Medicine</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anticonvulsant</td>
<td>3</td>
</tr>
<tr>
<td>Carbamazepine</td>
<td>3</td>
</tr>
<tr>
<td>Phenytoin</td>
<td>2</td>
</tr>
<tr>
<td>Antibiotics</td>
<td>3</td>
</tr>
<tr>
<td>Azithromycin</td>
<td>1</td>
</tr>
<tr>
<td>Cotrimoxazole</td>
<td>1</td>
</tr>
<tr>
<td>Non-steroidal anti-inflammatory</td>
<td>1</td>
</tr>
</tbody>
</table>

Table 2. Ocular complications in Steven-Johnson Syndrome

Ocular involvement was mild to moderate in 9 cases [Table II]. Lid complications were seen in 4 patients, lid edema with blisters [n=1], bullous lesion [n=2], meibomitis and blepharitis [n=1] with thick discharge on lid margin. None of the patients had dystrichiasis, trichiasis, entropion or ectropion. Conjunctival involvement was in the form of dry eye [n=5], congestion [n=1], membrane formation [n=1] and symblepharon [n=1]. Only one patient had corneal epithelial defect.
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MEMBRANE FORMATION IN SJS SYMBLEPHARON FORMATION IN SJS

<table>
<thead>
<tr>
<th>Ocular structure</th>
<th>Clinical findings</th>
<th>No of Patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lids</td>
<td>Oedema/thickening Bullous lesion Meibomitis Thick discharge Trichiasis Entropion/Ectropion Madarosis</td>
<td>1 2 1</td>
</tr>
<tr>
<td>Conjunctiva</td>
<td>Congestion Dry eye Symblepharon Membrane in the fornix</td>
<td>1 5 1 1</td>
</tr>
<tr>
<td>Cornea</td>
<td>Spk Epithelial defect Scarring Vascularisation Keratinisation Thinning</td>
<td>1</td>
</tr>
</tbody>
</table>

Treatment was started soon after diagnosis.
Suspected drug causing SJS was withdrawn.
Ocular management included frequent topical lubricants (Carboxymethyl cellulose with biodegradable preservative) in all patients. A topical antibiotic [usually tobramycin], was added where we suspected secondary infection.
Flurometholone was added for those with excessive ocular surface inflammation.
Membranolysis with glass rod passing was done 2-3 times daily for those who had symblepharon or pseudomembrane formation.
The ordinary thermometer serves well as the blunt glass rod.
All patients were encouraged to move the eyes voluntarily and to separate the lids frequently in order to prevent symblepharon formation.
Good ocular hygiene was ensured. Bandage contact lens was used in the patient with corneal epithelial defect.
All were put on systemic steroids; fluid and electrolytes were maintained. Ophthalmic followup was done once a day for each patient till they were discharged.
All patients were explained about the chronicity, severity and sequelae of the disease, possibility of dry eye and need of regular and long term ophthalmic follow up.
There was no mortality and all were discharged within [2 weeks to 2 months]. All patients were followed at 1 and 3 months after recovery of the acute phase.
9 patients recovered without any ocular sequelae. One patient had punctual stenosis, and minimal ankyloblepharon. One patient had inferior corneal opacity, which was not visually significant. Superficial punctate keratitis was seen in one patient at one month followup. All patients were advised to use lubricants for a long period.

Discussion:
SJS is an important cause of ocular morbidity. This analysis aimed to study the presenting features, possible etiologic factors and the outcome of intensive ocular management in reducing the sequelae. Acute phase includes lid oedema, blepharitis, conjunctivitis, pseudomembrane or membranous conjunctivitis. Later complications include from lid scarring entropion, ectropion, trichiasis, lagophthalmos, conjunctival scarring symblepharon or ankyloblepharon.
Tear film deficiency leads to conjunctival and corneal xerosis.
Late phase corneal complications develop due to corneal exposure leading to recurrent epithelial defect, corneal neovascularisation, conjunctivalisation of cornea, and corneal opacity. Ocular cicatricial pemphigoid has been reported 31 years after SJS in five patients.
Our analysis reconfirms that the most sight threatening sequelae can be prevented by aggressive medical management in the acute phase. Only one patient had a small corneal opacity and 5 patients had mild ocular surface disease. Corneal transplantation has poor visual prognosis with ocular surface disorders. It is also difficult to correct the structural abnormalities of lid & conjunctiva in the late phase as the tissues are fibrotic and friable. Most patients might require extensive surgical procedures like, amniotic membrane transplantation, skin graft etc. Osteooodontokeratoprosthesis is a recent innovative method to restore the vision of patients with endstage severe ocular surface disorder using autologous canine tooth and buccal mucosa as the artificial cornea or keratoprosthesis. Role of systemic corticosteroids in modulating ocular manifestations is not established. Cyclosporin A and plasmapheresis have been proposed as alternatives. Recently, IV immunoglobulin has also been advocated for both SJS

In contrast to many reports where Sulphonamides was the most common etiology, our patients had history of intake of anticonvulsants and other antibiotics. There is an immunologic susceptibility to the development of SJS. It has been reported that patients with HLA-Bw44 are more susceptible to SJS. Genetic factors are suspected, the suspected drug should not be used in the blood relatives of the patient. At this time the best result come from early diagnosis, immediate discontinuation of any suspected drug. Daily examination by an ophthalmologist is a must to prevent sight threatening sequelae.

References:


