Outcome of vigilant management of early Steven Johnsons Syndrome

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Abstract

Steven Johnsons Syndrome [SJS] is an important cause of ocular morbidity. We present a series of 14 SJS patients who presented to our institution in the acute phase. All patients with the diagnosis of SJS over last 2 years 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 fornice and voluntarily moving the eyes. This prevented formation of symblepharon, ankyloblepharon and other sequlae. All patients recovered without any sight threatening sequlae. Mild sequlae included - one patient had ankyloblepharon and punctual stenosis, another had inferior corneal opacity, mild dry eye was seen in 11 cases. This study lays emphasis on the importance of early aggressive management in preventing and reducing the severity of sight threatening sequlae of SJS.

Steven – Johnsons Syndrome [SJS] is a symptom complex characterized by symmetrically distributed erythematous 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 induced.1 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 cicatricial changes and perforation. Prompt diagnosis, identification and early withdrawal of all suspect drugs are the most important.2. The management of the patients must be undertaken in the specialized intensive care units. Ocular sequlae can be minimized by prompt, early and regular ophthalmic care.

We present a series of 14 patients [Aged 10-58 years with male: female ratio of 5:9] who presented to our institution in the last 2 years. All presented in the acute phase of SJS within 1 week of onset of symptoms. Eleven of these patients had ocular involvement. 3 of the patients first consulted an ophthalmologist, with only ocular complaints. All patients had a history of drug intake. Use of any systemic drug within 2-3 weeks of onset of prodromal symptoms was considered as possible etiological factor [Table-I]. In our series anticonvulsants were the most common etiologic factor especially - Carbamazepine [n=5] and Phenytoin [n=2].

Ocular involvement was mild to moderate in 11 cases [Table II], and 3 had no ocular involvement. Lid complications were seen in 5 patients, lid edema with blisters [n=1], madrosis [n=1], meibomitis and blepharitis [n=5] with thick discharge on lid margin. None of the patients had dystrichiasis, trichiasis, entropion or ectropion. Conjunctival involvement was in the form of congestion, membrane formation [n=3] and symblepharon [n=5]. Only one patient had corneal epithelial defect.
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, although the culture was negative. Fluorometholone 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 patients were put in isolation wards to prevent the chance of secondary infection and those requiring greater supportive measures were put in intensive care units. All were put on systemic steroids; fluid and electrolytes were maintained. Ophthalmic followup was done once or twice daily 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 2months. All patients were followed at 1 and 3 months after recovery of the acute phase. 11 patients recovered without any ocular sequelae. One patient had punctual stenosis, and minimal ankyloblepharon [Fig. 1]. 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

![Fig. 1. Child with ankyloblepharon, dry eye and conjunctival congestion](image-url)
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.4

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. Our 14- year old patient had the most severe ocular and systemic involvement. SJS in children is less common with ocular involvement in 39% cases in a 10-year series.5

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. Osteoodontokeratoprosthesis6 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.1

In contrast to many reports where Sulphonamides was the most common etiology, our patients had history of intake of anticonvulsants and other antibiotics. Carbamezepine was found to be the commonest cause of SJS in another report as well from Kerala.7 There is an immunologic susceptibility to the development of SJS. It has been reported that patients with HLA-Bw44 are more susceptible to SJS 8. 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, supportive therapy often in intensive care unit and paying close attention to ocular complications. Daily examination by an ophthalmologist is a must to prevent sight threatening sequelae.

References


