



OCULAR MANIFESTATIONS IN STEVENS JOHNSON SYNDROME PATIENTS ATTENDING TO GOVT.REGIONAL EYE HOSPITAL,KURNOOL.

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ABSTRACT **Aim:** To study the ocular manifestations in Stevens Johnson Syndrome
Methods: A total of 100 patients of all age groups with dermatological disorders attending to Regional Eye Hospital, Kurnool during the period June 2017 to June 2018 were examined and patients with features of Stevens Johnson syndrome were exclusively included in the study and examined which include visual acuity testing using Snellen chart, refraction, slit lamp biomicroscopy.
Results: Out of 100 patients, 15 patients were identified to have Stevens Johnson syndrome. 6 (40%) patients were between the age group 31-40. Males were 9(60%), females were 6 (40%). Bilateral involvement was seen in 10 (66.66%) cases.
Conclusion: In our study most common age group to be involved was between 31-40 years, males were more commonly involved than females, most common ocular manifestation was found to be conjunctivitis in 14(93.33%) patients.

KEYWORDS : Stevens Johnson Syndrome, lid edema, conjunctivitis, superficial punctate keratitis.

INTRODUCTION

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are hypersensitive mucocutaneous diseases that occur mostly by medication and infection. They have a direct effect on the skin, mucous membrane and eyes. Sometimes it can be life-threatening¹. SJS and TEN are variants belonging in the same class and are defined based on the amount of epidermal detachment. In SJS, 10% or less of the total body surface area is involved. In TEN, 30% or greater involvement is seen and SJS/TEN overlap between 10-30%.² Clinical findings include prodromal symptoms of fever and malaise, followed by the development of a generalized, tender cutaneous eruption consisting of macules, papules, atypical target lesions, and vesicles or bullae³. Serious dermatological manifestations make a physician overlook ocular sequelae, which are irreversible and fatal to visual acuity by the destruction of the ocular surface. The pathogenesis of SJS/TEN is controversial. A cytotoxic T lymphocyte (CTL) immune-mediated reaction is known as the major immunologic component of SJS/TEN^{4,5}. The ocular manifestation of SJS/TEN in various clinical stages; acute, subacute and chronic.

ACUTE PHASE

The pathogenesis of the acute phase is the keratinocyte apoptosis and secondary effects of inflammation and loss of ocular surface epithelium. Acute ocular involvement rate is 50% to 88% of SJS/TEN⁶⁻⁸. Epithelial loss of tarsal conjunctiva and eyelid margin with or without pseudomembrane or true membrane formation can make early symblepharon formation and fornix foreshortening. The corneal epithelial defect can bring out corneal ulceration and perforation⁹. Meibomitis has a high prevalence rate, seen in more than half of patients.¹⁰

SUBACUTE PHASE

Even though skin lesions are mostly resolved, chronic cicatrizing conjunctivitis with trichiasis and irregular eyelid margins may be persistent by the inflammation and ulceration of the ocular surface. The abnormal eyelid with misdirected and /or distichiasis lashes causes a mechanical abrasion on the corneal epithelium, leading to corneal epithelial defects, infection, and stromal scar. Severe inflammation and persistent ulceration of the tarsal conjunctiva and lid margins leads to lid margin keratinization and tarsal scar.^{11,12}

Sotozono and colleagues developed a grading system for the chronic ocular manifestations in patients with SJS/TEN which included corneal, conjunctival, and eyelid complications and they assessed 13

components and scored them on a scale from 0 to 3 according to their severity¹³. The grading system was based on manifestations observed using slit-lamp biomicroscopy. The classified corneal complications included superficial punctate keratopathy, epithelial defect, loss of the palisades of Vogt, conjunctivalization, neovascularization, opacification, and keratinization. The conjunctival complications include hyperemia and symblepharon formation. Eyelid complications include trichiasis, mucocutaneous junction involvement, meibomian gland involvement, and punctal occlusion.

The chronic ocular sequelae occur in up to 35% of SJS/TEN patients.¹⁴ Chronic ocular sequelae with severe visual loss is associated with lid margin abnormality and ocular surface failure.¹⁵ Conjunctival ulcerations or conjunctival membrane formation, or persistent inflammation makes permanent symblepharon and ankyloblepharon, which disrupts a tear film meniscus and inhibits proper eyelid closure and blinking¹⁵. The contracture of the palpebral conjunctiva leads to cicatricial entropion and trichiasis. Tarsal conjunctival scarring can be associated with eyelid malposition, ectropion, entropion, trichiasis, distichiasis, meibomian gland atrophy and inspissation, punctal occlusion, and keratinization of the eyelid margin, tarsal and bulbar conjunctival surfaces.¹⁶

MATERIALS AND METHODS

A total of 100 patients of all age groups with dermatological disorders attending to Regional Eye Hospital, Kurnool during the period June 2017 to June 2018 were examined and patients with features of Stevens Johnson syndrome were exclusively included in the study. Patients of all age groups with the involvement of at least one eye were included in the study. All the patients who were included in the study underwent visual acuity testing using a Snellen chart. Distance and near visual acuity, both presenting and best corrected after refraction, were measured for each eye separately using Snellen chart. Refraction is done. External eye examination, assessment of pupillary reaction, and anterior segment examination were done with slitlamp biomicroscope.

RESULTS

Table 1: distribution of age

Age	No of cases
0-10yrs	0(0%)
11-20yrs	3(20%)
21-30yrs	5(33.33%)
31-40yrs	6(40%)

41-50yrs	1(6.6%)
Total	15(100%)

Table 2: distribution of gender

Gender	No of cases
Males	9(60%)
Females	6(40%)

Table 3: laterality of eye involved

Laterality	No of cases
RE	2(13.33%)
LE	3(20%)
BE	10(66.66%)
Total	15(100%)

Table 4: ocular manifestations of Steven Johnsons Syndrome

Ocular manifestations	No of cases
Lid signs	
Lid edema	12(80%)
Ectropion	1(6.6%)
Entropion	1(6.6%)
Conjunctival signs	
Conjunctivitis	14(93.33%)
symblepharon	1(6.6%)
Corneal signs	
Superficial punctate keratitis	8(53.33%)
Epithelial defects	2(13.33%)

Fig 1:conjunctivitis and matting of eyelashes in SJS

DISCUSSION

Stevens Johnson Syndrome is an acute inflammatory polymorphic disease affecting the skin and mucous membranes. All ages may be affected and the incidence is equal in both sexes. This is a severe disease with a 5%-15% mortality rate. Ocular involvement, which occurs in as many as half patients, varies from mild mucopurulent conjunctivitis to severe perforating corneal ulcers. Blindness occasionally occurs in patients with severe late phase corneal complications such as ulceration, vascularization, and perforation.

In our study, 15 patients had manifestations of Stevens Johnson Syndrome out of 100 patients. Among them 3(20%) patients were between age group 11-20years, 5(33.33%) patients were between age group 21-30 years, 6 (40%) patients were between age group 31-40years, 1 (6.6%) patient between age group 41-50years.

In our study 9 (60%) were males and females were 6(40%)

In our study Right eye involvement was seen in 2 (13.33%) patients, left eye involvement was seen in 3 (20%) patients and both eyes were involved in 10 (66.66%) patients.

Among various ocular manifestations, the most common ocular structures to get involved were eyelids, conjunctiva, and cornea. Eyelid manifestations that were noticed include lid edema 80%, ectropion 6.6%, entropion 6.6%. Conjunctival manifestations that were noticed include conjunctivitis 93.33%, symblepharon 6.6%. corneal signs that were noticed include superficial punctate keratitis 53.33%, epithelial defects were 13.33%.

CONCLUSION

Ocular involvement in Stevens Johnson Syndrome begins with edema, erythema, and crusting of the eyelids. Concomitant conjunctivitis appears that is characterized by watery discharge with mucoid strands.

Secondary infection, with staphylococcus species may develop. In severe cases, membranous or pseudomembranous conjunctivitis may result along with symblepharon formation. Corneal involvement in the form of superficial punctate keratitis and epithelial defects may also be noted.

Late ocular complications include anomalies of the eyelid (ectropion, entropion) trichiasis and symblepharon.

Attentive examination and management in the acute stage provide the best opportunity to prevent chronic disease. There are changes in the ocular surface in every stage of the disease, when left untreated can become irreversible, and a lost opportunity to improve the visual function and quality of life of these patients.

REFERENCES

- Bastuji-Garin S, Fouchard N, Bertocchi M, Roujeau JC, Revuz J, Wolkenstein P. SCORTEN: a severity-of-illness score for toxic epidermal necrolysis. *Journal Of Investigative Dermatology*.2000;115:149-53.
- Bastuji-Garin S, Rzany B, Stern RS, Shear NH, Naldi L, Roujeau J. CLinical classification of cases of toxic epidermal necrolysis, Stevens-Johnson syndrome, and erythema multiforme. *Archives of Dermatology* 1993;129:92-6.
- Nickoloff BJ. Saving the skin from drug-induced detachment. *Natural Medicine*. 2008;14:1311-3.
- Kohanim S, Palioura S, Saeed HN, Akpek EK, Amesua G, Basu S, Blomquist PH, Bouchard CS, Dart JK, Gai X, Gomes JA. Stevens-Johnson syndrome/toxic epidermal necrolysis—a comprehensive review and guide to therapy. I. Systemic disease. The ocular surface. 2016 Jan 1;14(1):2-19.
- Friedmann PS, Strickland I, Pirmohamed M, Park B. Investigation of mechanisms in toxic epidermal necrolysis induced by carbamazepine. *Archives of Dermatology*. 1994;130:598-604
- Lopez-Garcia JS, Rivas Jara L, Garcia-Lozano CI, Conesa E, de Juan IE, Murube del Castillo J. Ocular features and histopathologic changes during follow-up of toxic epidermal necrolysis. *Ophthalmology*.2011;118:265-71.
- Power WJ, Ghorraishi M, Merayo-Lloves J, Neves RA, Foster CS. Analysis of the acute ophthalmic manifestations of the erythema multiforme/Stevens-Johnson syndrome/toxic epidermal necrolysis disease spectrum. *Ophthalmology* 1995;102:1669-76.
- Basu S, Pillai VS, Sangwan VS. Mucosal complications of modified osteo-odontokeratoprosthesis in chronic Stevens-Johnson syndrome. *American journal of ophthalmology*. 2013 Nov 1;156(5):867-73.
- Isawi H, Dhaliwal DK. Corneal melting and perforation in Stevens-Johnson syndrome following topical bromfenac use. *J Cataract Refract Surg* 2007;33:1644-6.
- Sachdev R, Bansal S, Sinha R, Sharma N, Titiyal JS. Bilateral microbial keratitis in highly active antiretroviral therapy-induced Stevens-Johnson syndrome and toxic epidermal necrolysis: a case series. *OculImmunolInflamm* 2011;19:343-5.
- Di Pascuale MA, Espana EM, Liu DT-S, Kawakita T, Li W, Gao YY, et al. Correlation of corneal complications with eyelid cicatricial pathologies in patients with Stevens-Johnson syndrome and toxic epidermal necrolysis syndrome. *Ophthalmology* 2005;112:904-12.
- Iyer G, Pillai VS, Srinivasan B, Guruswami S, Padmanabhan P. Mucous membrane grafting for lid margin keratinization in Stevens-Johnson syndrome: results. *Cornea* 2010;29:146-51.
- Sotozono C, Ang LP, Koizumi N, Higashihara H, Ueta M, Inatomi T, et al. New grading system for the evaluation of chronic ocular manifestations in patients with Stevens-Johnson syndrome. *Ophthalmology* 2007; 114:1294-302.
- Sotozono C, Ueta M, Koizumi N, Inatomi T, Shirakata Y, Ikezawa Z, et al. Diagnosis and treatment of Stevens-Johnson syndrome and toxic epidermal necrolysis with ocular complications. *Ophthalmology* 2009;116:685-90.
- Jain R, Sharma N, Basu S, Iyer G, Ueda M, Sotozono C, Kannabiran C, Rathi VM, Gupta N, Kinoshita S, Gomes JA. Stevens-Johnson syndrome: The role of an ophthalmologist. *Survey of ophthalmology*. 2016 Jul 1;61(4):369-99.
- Tseng SC, Di Pascuale MA, Liu DT, Gao YY, Baradaran-Rafii A. Intraoperative mitomycin C and amniotic membrane transplantation for fornix reconstruction in severe cicatricial ocular surface diseases. *Ophthalmology* 2005; 112:896-903.