Case Report

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# Lacrimal Drainage System Involvement in Stevens-Johnson Syndrome: A Case Report and Literature Review

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## ABSTRACT

Stevens-Johnson-Syndrome (SJS) is characterized by severe inflammation of the skin and mucous membranes of the entire body. Also known as Lyell disease, the effect of the disease is usually quite fatal if not aggressively managed. The lacrimal drainage system which is lined inside by the mucous membrane is also not spared and when involved the management is challenging. We report a 31-year-oldretroviral disease positive female on antiretroviral drugs whose consent was sought and permission granted. She presented with severe symblepharon occluding the puncta of the upper and lower lids, associated with persistent epiphora and bacterial blepharitis in both eyes, with good visual acuities and multiforme erythematous skin eruptions. The probing of the system revealed soft stops along the canalicular and naso-lacrimal duct segments of the lacrimal drainage systems. There was failed venflon stenting procedure after symblepharon release warranting bilateral dacryocystorhinostomies (DCRs) with silicone intubations which was followed by marked relief of symptoms and signs. We therefore conclude that SJS can affect the lacrimal drainage systems and the management challenges are enormous requiring a lotof resilience and patience on the side of the oculoplastic surgeon.

**Keywords:** Stevens-Johnson Syndrome, Lacrimal Drainage System, Epiphora, Dacryocystorhinostomy Silicone Intubation.

## INTRODUCTION

Stevens- Johnson Syndrome is an immune-complex mediated hypersensitivity reaction involving the skin and mucous membrane of the whole body. It varies from mild skin and mucous eruptions to very severe and sometimes fatal systemic illness. Ophthalmic Stevens-Johnson syndrome can be very severe leading to a number of morbidities involving both ocular and ocular adnexal tissues resulting even to blindness.

### Case Report

A 31year old lady presented to us with a 6-months history of epiphora and persistent itching and redness in both eyes. She volunteered a positive history of Retroviral disease on Highly Active Antiretroviral Therapy (HAART). Her general examination revealed multiple hyperpigmented skin lesions almost covering

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the entire body. Her eye examination revealed unaided visual acuities of 6/9 in both eyes. Her intra-ocular pressures were within normal. The lid examination revealed epiphora, blepharitis and symblepharon connecting the upper and lower puncta of both eyes (Figures.1&2).


Fig. 1


Fig. 2

Her laboratory investigations did not reveal any abnormalities except that she tested positive for Human Immunodeficiency Virus(HIV) using the Enzyme Linked Immunosorbent Assay(ELISA)screening test. The symblepharon were released and upper and lower punctoplasties successfully carried out. Soft stops were encountered mid –canalicular in the upper and lower canaliculi of the two eyes but were overcome with gentle force during naso-lacrimal duct probing using the bowman lacrimal probes, suggesting partial blockages of all the canaliculi of both eyes. Resistance was also encountered in both the naso-lacrimal ducts but was also overcome by gentle push of the probes. Syringing with normal saline was done and only mild regurgitation of the fluid in the upper puncta, suggesting the partial blockage in the naso-lacrimal duct. Venflon intubations were carried out successfully in the lower canaliculi. The patient was discharged home a day after the surgical interventions. After some three months the patient reported back with complaints of stent or venflon tube extrusion associated with mild epiphora. The tube reinsertion was attempted but failed. Patient was readmitted and a more definitive surgery(DCR with intubation) was carried out successfully under general anaesthesia. The patient was discharged on the 3rd day post surgery (See Figures 3&4)

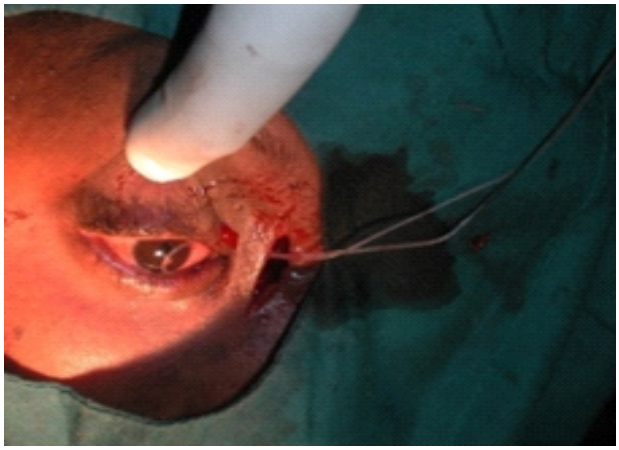
Fig. 3

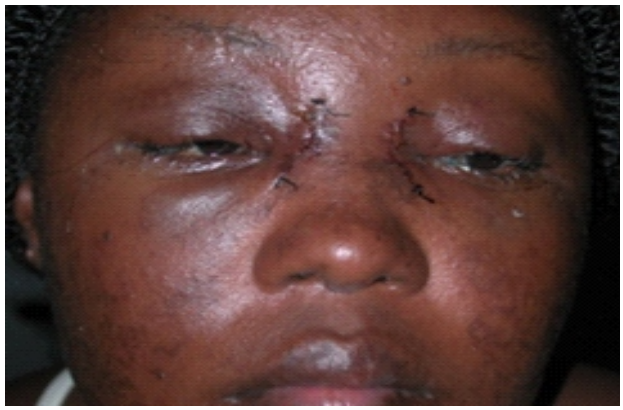
Fig. 4

There was marked improvement of symptoms with all tubes in situ after three months of follow-up. (See Fig. 5)

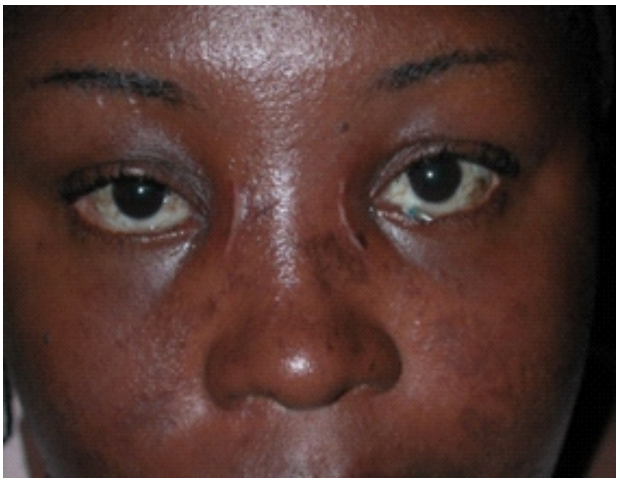
Fig. 5

## DISCUSSION AND LITERATURE REVIEW

Stevens-Johnson Syndrome was first described in 1922 by two pediatricians by the names: A.M. Stevens and F.C.Johnson. They reported 2 cases of boys from the same neighborhood in New York City whose presentations were similar with febrile illness and mucocutaneous eruptions involving the mouth and the eyes (muco-purulent discharges from the mouth and the eyes). Stevens - Johnson syndrome was also known as the Lyell Syndromeas their presentation was initially misdiagnosed as cases of hemorrhagic measles or black measles until the two pediatricians discovered that these were different entities morphologically[1,7]. The more severe variant of the syndrome is referred to as Toxic Epidermal Necrolysis (TEN) and it is characterized by intense life-threatening blistering and necrosisof the skin requiring intensive care of the severe pains, multiple blisters, respiratory distress and dehydration[2].

Together with TEN, SJS affects 1 to 2 people per million per year. Typical onset is under the age of 30. It is believed to be a type IV hypersensitivity reaction and classed together with the drug reaction with eosinophilia and systemic symptoms (DRESS syndrome), acute generalized exanthematous pustulosis (AGEP) and toxic epidermal necrolysis (TEN) in a group of conditions known as severe cutaneous adverse reactions (SCARs).

Most common devastating complications of the syndrome are ophthalmic in nature with most severe sequelae on the ocular surface thus making the role of the ophthalmologist key in the management of SJS[2,10,11]. Our case had both the ocular and adnexal involvement with symblepharon and naso-lacrimal drainage system,but corneal- sparing complications.

Work done by Chie Sotozono, et al revealed loss of the palisades of Vogt (114 eyes; 82.6%) and meibomian gland involvement (102 eyes; 73.9%). Visual acuity in 74 of the 138 eyes (53.6%) was worse than 6/60[3]. Dry eye syndrome was one of the most common delayed complications of the disease in some studies and worse delayed complications were related to the severity of the acute phase of the disease [3,16]. Other ocular complications include conjunctival epithelial defects, pseudomembranous conjunctivitis, corneal neovascularization (pannus formation) with resultant corneal blindness [4,12]. The adnexal involvement is also fairly common with cicatricial ectropion, entropion and partial or complete ankyloblepharon[3,4,12]. Our patient had symblepharon with punctal occlusions.

The major cause of Stevens-Johnson Syndrome is not known. The triggering factors for the syndrome can be classified into hereditary factors, drug-induced factors, and infections-induced factors. The drug-induced factors seem to be the most common trigger factors of the syndrome [5,8]. The drugs incriminated include: Anti-bacterials such as the sulphonamides and penicillins; Anti-virals such as Nevirapine (Viramune); Anti-fungals such as Fluconazole; Anti-inflammatories such as the Salicylates including; Acetaminophen, Ibuprofen and Naproxen sodium; Anti-convulsants such as barbiturates including phenobarbitone, barbital, phentoin and methohexital; Anti-psychotics like Lamatrigine; Anti-gout like Allopurinol and even Radiotherapy [5,9]

Infections-induced trigger factors implicated include the Human Immune Virus (HIV), Mycoplasma pneumonia, Streptococcus, Meningococus and Hepatitis A virus [7,8]. Organ transplant and some blood cancers have been implicated as well[7,8].

Stevens-Johnson Syndrome is classified into 3, basically based on the degree of skin involvement as follows:

TYPICAL SJS: When less than 10 percent of the skin is involved.

INTERMEDIATE SJS: When between 10 and 30 percent of the skin is involved.

TEN: When more than 30 percent of the skin is involved.

Treatment of SJS/TEN is complex but basically supportive requiring hospitalization particularly in an intensive care unit and preferably in the burns unit. The main aim is to rehydrate, control the pains and minimize structural damage. So, pain medications to prevent pains, antibiotics to prevent infections, antihistamines to prevent secretions and itching, intravenous fluids to rehydrate, intravenous immunoglobulins and corticosteroids to modulate the immune systems and reduce structural damage. Yayoi Araki, et al found use of steroid pulsed therapy at the onset of the disease with topical bethamethasone very useful in the prevention of ocular complications[4]. Sterilised banana have also been found useful in the control of pains in some literatures [17]. Surgery comes handy in certain cases particularly where structural damages have occurred. In this case that we reported, there was some damage to the conjunctiva with symblepharon tagging the upper and lower puncta causing punctal blockage with epiphora, requiring surgical intervention. There were also canalicular and naso-lacrimal ducts obstructions requiring dacryocystorhinostomy with intubation which was successfully carried out.

Complications of SJS/TEN are multi-system involving, with varying prognosis. Almost all systems with mucosal linings in the body are vulnerable. In our case report, the ocular and adnexal structures were more affected, extending from the eye ball to the lacrimal drainage system requiring surgical interventions.

Skin lesions usually regrow over two to three weeks however, complete recovery can take months. The risk of death with SJS is 5 to 10%.

Diagnosis is confirmed by the presence of Nikolsky sign which is a skin finding in which the top layers of the skin slip away from the lower layers when rubbed[13]. Skin biopsy is helpful in the diagnosis of TEN.

## PROGNOSIS

Generally the outcome of SJS/TEN includes organ damage and even failure, ocular morbidity/ blindness and restrictive lung disease. Patients with SJS or TEN caused by drugs have better prognosis the earlier the causative drug is withdrawn [14].

The outcome of the SJS/TEN depends on the SCORTEN (the severity-of- illness score for TEN) scale[14,15,17]

**Severity-of-Illness Score for Toxic Epidermal Necrolysis (SCORTEN)**

| Risk Factor* | 0 | 1 |
|---|---|---|
| Age | < 40 years | ≥ 40 years |
| Associated cancer | No | Yes |
| Heart rate (beats/minute) | < 120 | ≥ 120 |
| Serum blood urea nitrogen | ≤ 28 mg/dL (10 mmol/L) | > 28 mg/dL (10 mmol/L) |
| Detached or compromised body surface | < 10% | ≥ 10% |
| Serum bicarbonate | ≥ 20 mEq/L (≥ 20 mmol/L) | < 20 mEq/L (< 20 mmol/L) |
| Serum glucose | ≤ 250 mg/dL (≤ 13.88 mmol/L) | > 250 mg/dL (> 13.88 mmol/L) |

** More risk factors indicate a higher score and a higher mortality rate (%) as follows*

- *0 -1 =3.2% (CI: 0.1 to 16.7)*
- *2 = 12.1% (CI: 5.4 to 22.5)*
- *3 = 35.3% (CI: 19.8 to 53.5)*
- *4 = 58.3% (CI: 36.6 to 77.9)*
- *≥ 5 = > 90% (CI: 55.5 to 99.8)*

*CI= confidence interval.*

## CONCLUSION

SJS/TEN can affect virtually all systems lined by mucous membrane causing serious morbidity and even death. In our case report there were serious ocular and adnexal structural complications warranting surgical intervention. Immediate intervention will reduce the rate and severity of complications. We therefore

recommend that where epiphora presents, whether during the active or chronic stages of SJS/TEN, the ophthalmologist should do a thorough assessment of the lacrimal drainage system. Where there is no ophthalmologist or better still, an oculoplastic surgeon, then the patient should be referred accordingly.

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