Case Report

Preventable Blindness due to Ankyloblepharonin Stevens-Johnson Syndrome–A Case Report

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Abstract

Stevens-Johnson syndrome (SJS) is common, having long term ocular complications ranging from dry eye to loss of vision due to ankyloblepharon or corneal opacities. Proper medical management and separating the bulbar conjunctiva from palpebral regularly by a glass rod during the acute phaseof the diseases can prevent the development of ankyloblepharonand symblepharon. Here we present a case of ankyloblepharon after SJS which was repaired after 6 months in the department of ophthalmology, BIRDEM General Hospital.

Key word:ankyloblepharon; Stevens-Johnson syndrome;symblepharon

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Introduction

Stevens-Johnson syndrome(SJS) causes various ocular complications in which symblepharon and ankyloblepharon are very common. Ankyloblepharon is potentially blinding, which can be prevented by adequate medical measurement. Here we present such a case.

Case Report

A 16-year-old girl presented with complete obscuration of vision and fusion of eye lids of both eyes for the last six months. About sixmonths back she developed diffuse rash in her face and body after taking an analgesic. The rash spread throughout allover the body including limbs, trunk and face. The eyes and oral cavity were also involved. She was diagnosed as a case of SJS in a tertiary hospital and was treated conservatively with systemic antibiotic and steroids. Ophthalmic drops were prescribed, but not instilled properly and no separation of bulbar and palpebral conjunctiva was done. The eyes remained closed. After a few weeks, she got well but the eyelids remained fused with each other and the patient became blind.

At our care, on ocular examination, both eyes were found to have complete ankyloblepharon having no vision in both eyes. Behind the fused eyelids the movement of eyeballs were sufficient. The contour of the eyeballs were also normal (Figure 1). General examination reveals diffuse skin pigmentation in the face and body (Figure 2).The patient was diagnosed as a case of bilateral ankylobleaphron due to SJS.

The patient underwent lid reconstruction surgery (separation of fused eyelids, separation of adhesion between bulberand palpebral conjunctiva) and contact shield was placed on both eyes.On the first postoperative day, the patient gained formed vision (right eye 6/60 and left eye 6/24).Postoperatively she was treated with topical antibiotic, steroid and artificial tear.

Gradually the vision of the patent improved but ocular surface was dry.Cornea of both eyes werevascularised from the periphery to midperiphery.

After one month, the patent gained vision up to 6/12 in both eyes (Figure 3).Now the patient is getting topical artificial tears frequently and is being observed in outpatient department regularly at an interval of three months.

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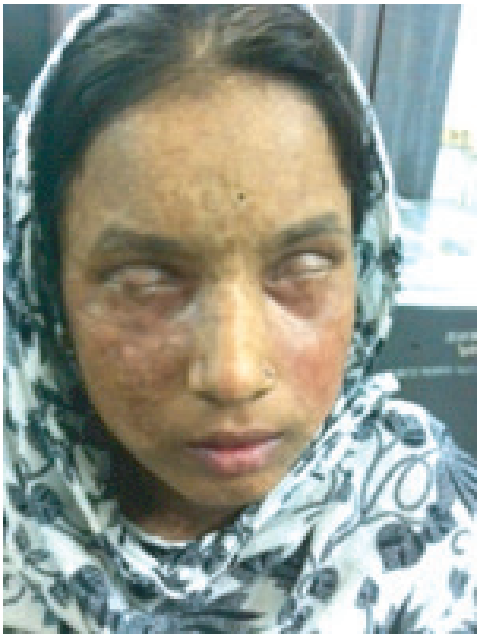

Figure-1: *Total ankyloblepharon at presentation*

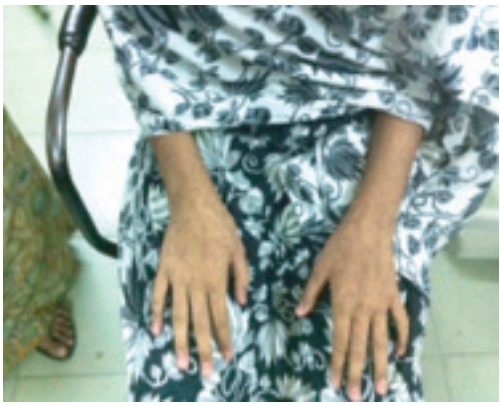

Figure-2: *Skin pigmentation*

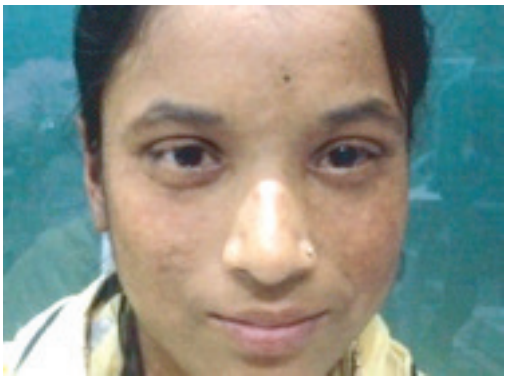

Figure-3: *After 1 month of lid re construction surgery*

Discussion

SJS and toxic epidermal necrolysis (TEN) are conditions characterized by intra-epidermal cell death leading to diffuse vesicobullous eruptions. The differentiating criteria for SJS and TEN is the extent of skin detachment SJS is defined as <10% total body surface area, SJS-TEN overlap as 10-30% and TEN as >30%. Mucosal involvement is a common, occurring in up to 90% of patients and may involve the ocular surface in as many as 80% of patients.[1] The incidence of SIS/TEN is low, estimated as 1–7 cases per million per year.[2]

The exact pathogenesis of SJS/TEN is unknown but appears to involve cell-mediated keratinocyte apoptosis via the Fas signaling cascade and granulosin.[3] The syndrome can result from exposure to certain medications, infections or malignancy, though almost a quarter of cases have no trigger.[4] Medications are the most frequently implicated inciting factor with antibacterial sulfonamides, such as trimethoprim/ sulfamethoxazol, and anticonvlsants, such as phenytion, as the leading culprits. Infections are the next most common cause. There is strong association with *Mycoplasma pneumoniae*in children, but other infectious causes of SJS/TEN are relatively rare. In addition, it is important to note that HIV patients have up to a hundred-fold increase in susceptibility, likely due to immunologic abnormalities and intensive drug regimens. Many other medications and infectious agents have been associated with SJS/TEN, but the most common etiologies are listed pharmacological (anticonvulsants, carbamazepine, NAIDS etc), infectious (Herpes simplex virus, Cytomegalovirus) etc.[2]

Amongst survivors, long-term ocular complications can be serious and are thought to affect approximately 60% of patients.[6]Corneal damage in the form of scarring or limbal stem cell failure is the most severe ocular outcome.Conjunctival scarring can contribute to long-term corneal pathology and subsequent visual impairment.For example, palpebral conjunctival scarring can cause chronic micro-trauma with the blink reflex, while symblepharon formation can lead to poor tear film dynamics and predisposition to severe dry eye.

Because ocular involvement is common and there is potential for severe visual consequences, all patients with SJS/TEN should be urgently evaluated by an

ophthalmologist. Fluorescein staining should be used to evaluate the extent of corneal involvement and as it is frequently affected by SJS/TEN.

While systemic corticosteroids are frequently used for the management of SJS/TEN, widespread acceptance of this approach has long been controversial. Evidence for its efficacy is lacking and early studies associated with systemic steroids showed a slight increase in the mortality of pediatric patients. Intravenous-immunoglobulin (IVIG) administered with the goal of inhabiting the Fas-Ligand signaling pathway, has recently gained traction as a possible therapy but studies regarding its efficacy continue to have conflicting findings. Of note, one recent study found improved ocular outcomes associated IVIG therapy compared to systemic steroids.[9]

Studies investigating the therapeutic value of topical medications of ocular SJS/TEN are similarly lacking. While there is no standard treatment, a combination of topical corticosteroids and antibiotics are often used in cases of mild ocular involvement, with one retrospective study suggesting that early topical steroids are associated with improved visual outcomes.[10] For more server ocular involvement, there is evidence that early surgical intervention with amniotic membrane can lead to improved outcomes.[7,11,12]

In this particular case, during the acute attack, ocular management was not proper. Ocular medications were instilled outside of the eye as because the patient didnot allow separating the eyelids from each other due to pain. Thus gradually the eyelids were adhered with each other permanently and developed total bilateral ankyloblepharon.

Conclusion

SJS can cause severe ocular complication like total ankyloblepharon which can lead to total blindness. Proper medical management and regular separation of bulbar and palpebral conjunctiva by a glass road during the acute stage of the diseases can prevent such complications.

Conflict of interest: None.

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