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Bilateral Levator Aponeurosis Disinsertion Following Stevens – Johnson Syndrome: Case Report

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Abstract **Case Report**

Stevens–Johnson syndrome is complex immunological syndrome characterized by acute blistering of skin and mucous membrane. The disease frequently involves the eye and can cause blindness. Here we report an unusual occurrence of bilateral blepharoptosis due to bilateral levator aponeurosis dehiscence following Stevens –Johnson's syndrome.
Key words: Stevens – Johnson, syndrome, levator aponeurosis, dehiscence.

INTRODUCTION

Stevens–Johnson syndrome (SJS), also known as erythema multiforme major is a complex immunological syndrome characterized by acute blistering affecting the skin and at least two mucous membranes. Toxic epidermal necrolysis (TEN) is the most severe form of erythema multiforme and involves greater than 30% of the skin, which sloughs in sheets [1]. Stevens – Johnson syndrome frequently involves the eye and in severe cases can result in blindness. The acute manifestation of the disease on ocular structures (eyelids, conjunctiva and cornea) occur in 60 to 100 percent cases while the late complications have been reported to occur in 20 to 80% of survivors [2, 3].

A recent literature search by us revealed no reported case of levator aponeurosis dehiscence associated with Steven Johnson syndrome.

Here we report an unusual case of bilateral blepharoptosis due to levator aponeurosis disinsertion following Stevens –Johnson syndrome.

CASE REPORT

A 27 year old female presented to our hospital with 7 years history of drooping of both eyelids. The droop was preceded by a history generalized mucocutaneous rashes that developed suddenly within 2 weeks of ingestion of an anti-malaria drug containing sulfadoxine and pyrimethamine (figure 1). She was treated then at a peripheral hospital where she was told she had Stevens-Johnson syndrome. She also complained of persistent loss of vision and burning sensation on both eyes and foreign body sensation on the right eye following the above incident. However her major concern was the droopy eyelids. The burning sensation was relieved by the use of artificial tears.

Examination revealed visual acuity of 1/60 on the right eye and 6/60 on the left eye. There was bilateral ptosis with high upper eyelid crease. Margin reflex distance (MRD 1) was -4mm and -3mm on the right and left eye respectively and levator function was 6mm and 7mm on the right and left eye respectively. The right eye was exodeviated and there was entropion of the right upper eyelid with trichiasis. Extraocular muscle motility in both eyes was full in all directions of gaze. The conjunctiva of both eyes was mildly hyperemic with evidence of xerosis. Both cornea were scarred with punctuate keratitis worse on the right eye. There was bilateral poor red reflex on fundoscopy worse on the right eye. Examination of photograph she took shortly before she suffered SJS revealed normal eyelids (figure 1). Diagnoses made were bilateral levator aponeurotic dehiscence and cornea scarring and right upper eyelid cicatricial entropion post Steven Johnson syndrome.

Routine hematological and biochemical tests done on her were normal. Screening for human immunodeficiency virus (HIV) was negative.

She underwent entropion surgery on the right eye and bilateral ptosis surgery in our centre. The levator aponeurosis was found to be disinserted from the anterior surface of the tarsus which appeared scarred in both eyes and was repositioned with good results (figure 2).

Fig-1: (A) Picture of patient showing skin rash that developed within 2 weeks of ingesting anti-malarial medication (B) Patient picture before the incident showing normal eyelids (C) Bilateral ptosis with high upper eyelid crease (D) Appearance of right eye after surgery and left eye before surgery with the degree of ptosis fully unmasked

Fig-2: Patient at 6 months post surgery with good position of upper eyelids

DISCUSSION

The pathophysiological mechanism of SJS is not fully understood. The disease is thought to be either a delayed hypersensitivity reaction to certain medications or a response to epithelial cell antigens modified by drug exposure. Genetic predisposition may also play a part due to a genetically determined enzyme deficiency for the metabolites of certain medicines [4]. The histopathological hallmark of this disease is wide-spread epidermal necrosis due to death by apoptosis of keratinocytes. CD8 cells act as mediators in this process [5]. The massive keratinocyte apoptosis renders the epidermis and epithelial surfaces disconnected from their vascular bed with resultant necrosis, inflammation, and sloughing [2, 5].

Drugs, infections and malignancies are recognized as triggers of SJS [5]. However, drugs are considered to be the main etiologic agent (50 to 80% of cases) and identified drugs include sulfonamide derivatives, anticonvulsants such as phenytoin, carbamazepine, Non-steroidal anti-inflammatory drugs, oxide inhibitors such as allopurinol etc [6]. Common infectious agent recognized to have a role in SJS are mycoplasma pneumoniae, enterovirus, hepatitis B virus, group A streptococcus and mycobacterium tuberculosis etc [7].

The reported acute manifestation of SJS is broad. In the acute phase of which usually occur within 2 weeks of onset of symptoms is marked by bilateral conjunctivitis, hyperemia, conjunctival membrane or pseudomembrane formation, meibomitis, symblepharon, conjunctival sloughing and cornea epithelia defects [2, 8]. The sub-acute stage of the disease is described as stage of smoldering chronic conjunctivitis with lid margin changes and trichiasis. At this stage most of the skin lesions are resolved [2]. Ophthalmic findings may include entropion, trichiasis, distichiasis, symblepharon formation, recurrent or persistent corneal epithelial defects, severe dry eye and keratinizaton of the posterior lid margin [8].

The chronic stage of the SJS is characterized by persistent and prolonged ocular surface inflammation and ulceration. Chronic ocular sequelae with severe visual loss are associated with lid margin abnormality and ocular surface failure [9]. Conjunctival ulcerations or conjunctival membrane formation as well as persistent inflammation can result in permanent symblepharon and ankyloblepharon, which disrupts tear film meniscus and inhibits proper eyelid closure and blinking, and sometimes restrict ocular motility [8]. There may be scarring with contracture of the tarsal conjunctiva causing malpositions of the eyelid and other disorders, including ectropion, entropion, trichiasis, distichiasis, meibomian gland atrophy and inspissation, punctal occlusion, and keratinization of the eyelid margin, tarsal and bulbar conjunctival surfaces, cornea opacification, vascularization or ulceration [2, 8].

The occurrence of bilateral blepharoptosis due to levator aponeurosis disinsertion in our patient is an unusual manifestation of SJS report of which was not seen in our literature search. The mechanism of the ptosis here is probably due to chronic inflammation induced stretching of the levator aponeurosis and

dehiscence from the anterior surface of the tarsal plate [10]. The chronic inflammation may account for the scarring of the anterior surface of the tarsus in both eyes seen in this patient.

The goal of treatment in SJS is survival and recovery from the systemic disease as well as the prevention of cicatricial complications in the affected organ systems [2].

The major aim of early ophthalmologic intervention in the acute stage of SJS is to prevent cicatricial complication and in late stages of the disease is reconstruction of ocular surface to correct the effects chronic inflammation. The management of late sequelae remains a challenge because of the irreversible alteration in the ocular surface [6].

In the acute stage, liberal use of lubricants (preferably non preserved) is the main stay of care. Prophylactic antibiotic and topical steroids have been reported to improve the outcome of care and have become an acceptable practice. Topical steroid drops plus ointment for the lid margin is recommended only after microbial keratitis has been excluded. A symblepharon ring can be used with copious lubrication to prevent adhesions in the fornix [2, 8].

Various surgical options are available for care of patients with SJS depending on the nature of ophthalmic complications. Mucus membrane transplantation and amniotic membrane grafting are increasingly being used to mitigate complications and the reconstruction the ocular surface in patients with SJS. Amniotic membrane tends to reduce ocular surface inflammation, form a scaffold for re-epithelialization, and prevent symblepharon formation, thus mitigating the long term sequelae of the disease [2, 11].

CONCLUSION

Stevens-Johnson syndrome is a complex immunological syndrome that can result in devastating ocular surface scarring and keratinization. This report adds to the spectrum of long term ocular complications associated with SJS. Controlling inflammation in the acute and late phases of the disease should help in reducing the occurrence of this unusual complication.

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