



Bilateral Conjunctival Inclusion Cysts as a Sequele of Stevens Jhonson's Syndrome-A Rare Case and Its Management

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Introduction

Steven Jhonson Syndrome is a major cause of non auto-immune cicatrizing conjunctivitis. It is a complex immunological syndrome characterized by blistering of skin and mucous membrane with female preponderance within 10-30 yrs. Etiology being infections, drug induced, malignancy and idiopathic (20-50%). Idiosyncratic, delayed hypersensitivity reaction leading to extensive surface denudation with proliferation of sequestered epithelium leading to inclusion cyst formation. Complications include ankyloblepharon, symblepharon, inclusion cyst, keratinisation, opacity of cornea, ectropion, entropion.

Case Report

A 21 year old female patient with SJS was referred to our hospital for evaluation of swelling of both lids of both eyes. She referred that the masses have been growing for 2 months. 3 years back she was hospitalized for SJS after consumption of sulfa drugs followed by attack of fever and skin rashes with ocular involvement.

On examination best corrected visual acuity was 6/9(RE) & 6/12(LE). Biomicroscopy revealed ovoid, translucent, cystic swelling in the lower fornix associated with ankyloblepharon and symblepharon. Corneal transparency was lost in lower third in LE. Anterior chamber was normal. Intraocular pressure was 14.6 mmhg in each eye. Fundus exam was normal with patent lacrimal passage. Normal tear film break up time with adequate tear meniscus.

She was treated by marsupialization of the cysts in two setting of alternate eyes. Amniotic membrane was used to cover the defect, sutured with vicryl 8'0.

Histopathological analysis revealed B/L inclusion cysts having mucoid filled spaces, subepithelial plasma cells, helper T cells. Margins were negative for metastatic changes. The systemic examination was negative for metastatic disease. Moxifloxacin (0.5%) with Dexamethasone (0.1%) eye drop was administered 6 times daily with tapering doses over 2 months for both eyes. She was followed up after 1, 4, 8 weeks post operatively with finding of residual entropion of lower lid of LE. Epilation was done for the same.

Discussion

Steven Johnson Syndrome (SJS) or Toxic Epidermal Necrolysis (TEN) is a life threatening immune complex mediated hypersensitivity reaction which mainly involves skin and mucous membrane. SJS is generally induced by the drugs. SJS predominately involves the oral mucosa and conjunctiva. Conjunctival involvement varies from 45% to 80%. Incidence being equal in both sexes. Clinical manifestations range from mild to severe. A prodrome of chills is followed by headache, pharyngitis, tachypnoea and tachycardia. Bullous mucosal lesions develop, especially in oropharynx. These lesions get rupture and ulcerate and covered by gray white membrane and haemorrhagic crust.

Detailed slit lamp evaluation includes lid pathologies like lid thickening, discharge, meibomitis, trichiasis, blepharitis, entropion and ectropion. Conjunctival pathologies included superficial punctuate keratitis, dry eye, thinning, scarring, vascularisation, infective keratitis and keratinization. Blindness occurs due to corneal complications in late phase.

Acute medical intervention involves both local and systemic measures for care of lids, conjunctiva and cornea. Medical therapy aims at reducing the sequelae of cicatrization with topical tear substitute, topical corticosteroids or topical broad spectrum antibiotics either alone or in combination. A dermatologist should be consulted. Ocular lubrication with artificial tears and ointments (preferably preservative free) should be applied regularly. Under topical anaesthesia, superior and inferior fornices should be inspected and debrided daily. A glass rod can be used for lysis of symblepharon. A symblepharon ring may be useful in severe cases in cooperative patients.

Amniotic membrane is the innermost layer of placenta and comprised of a thick basement membrane with a poorly cellularised stromal matrix. Its ability to enhance epithelisation, reduction of inflammation, inhibition of neovascularisation and prevention of ocular

surface scarring makes its use in ophthalmologic surgery fruitful. It delivers a complex signaling milieu consisting of immunomodulators and growth factors and provides substrate for epithelial cell growth. It may be used as a temporary bandage for permanent graft. This technique is generally reserved for patients with moderate or severe conjunctival involvement as these patients are more susceptible for visual loss from ocular surface scarring.

Conclusion

To our knowledge, this is the first reported case of SJS with ocular manifestation as inclusion cysts in our setup. These patients need periodic skin examination by dermatologists and ocular examination by an ophthalmologist. Patient need to avoid drug like sulfa drugs, analgesics, antiepileptics and penicillin to which they are most susceptible.

Declaration of Patient Consent: The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in journal. The patients understand that her name and essential records will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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