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Corneal Pathologies in Stevens Johnson Syndrome -An Ayurvedic Purview

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ABSTRACT

Stevens-Johnson syndrome (SJS) is a rare life-threatening hypersensitive mucocutaneous disease triggered mostly by medication and other diseases. Even after recovering from skin related issues, most of them are suffer from serious ocular complications leading to blindness. There is no definite effective treatment for SJS. Early detection and aggressive treatment are important for the improvement of the eye. Eyelid margin, palpebral conjunctiva and fornix should be checked properly to detect the cicatricial changes that make chronic ocular surface problems such as limbal cell deficiency and complete ocular surface keratinization. Hence, there is a need for identifying corneal pathologies as early as possible, to manage the later stages of disease and to reduce the risk of complication. In this presentation an effort has been made to review corneal pathologies in Stevens -Johnson Syndrome and made to correlate ayurvedic and modern corneal disorders in compliment aspect.

Key words: Stevens-Johnson syndrome (SJS), corneal pathologies, Savrana Shukla, Shushkakshipaka.

INTRODUCTION

Eye diseases affecting the cornea are a major cause of blindness worldwide. [1] According to the World Health Organization, corneal diseases are the major causes of vision impairment and blindness in the world nowadays, after cataract and glaucoma. In India, it is estimated that there are approximately 6.8 million people who have vision loss due to corneal diseases; of these, about a million have both eyes involvement. Systemic diseases affecting the cornea have a large range of manifestations. [2] Stevens-Johnson syndrome

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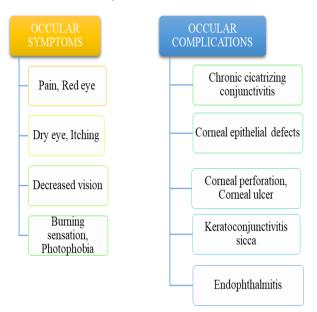
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(SJS) is a dermatologic emergency, characterized by the presence of epidermal and mucosal bullous lesions involving body surface. The accurate pathophysiology of Stevens-Johnson Syndrome is unspecified. Majority of SJS cases are attributed to delayed drug hypersensitivity reactions to a medication or medication metabolite.^[3] The severe cutaneous manifestations of this disease range may often lead to various types of the ocular seguelae, which are very common and may lead to visual damage. Most patients consult dermatologist or physician for skin lesions in the acute phase and consult ophthalmologist after recovering from lesions.[4] Hence, there is a need for identifying corneal pathologies as early as possible, to manage the later stages of disease and to reduce the risk of complication. The conforming behaviour of corneal disorders of Ayurveda and modern is justified by the clinical symptomology, progression of the disease, prognostic reasons, and relevant intervention. integrated disease approach, application and predicting prognosis will satisfy the resultant outcome of the disease in a more favourable

view which will augur a better perspective in managing preventable corneal blindness.

Stevens Johnson Syndrome - Occular Manifestations



Surgical correction may be hired to treat ocular sequalae; however, the results are mostly poor in terms of visual improvement and quality of life. Permanent visual impairment and ocular discomfort may continue throughout life, and patients may require lifelong medication and ophthalmic care. [5]

Corneal Pathologies in Stevens Johnson Syndrome

The following corneal pathologies and their ayurvedic aspects are studied in this article.

Keratoconjunctivitis Sicca (KCS)

Dry eye occurs when there is insufficient tear volume, ensuing in an unstable tear film and ocular surface complications. The common ocular symptoms are feelings of dryness, grittiness and burning that specifically exacerbate over the course of the day. It is an extremely familiar condition, especially in postmenopausal women and the elderly. Keratoconjunctivitis sicca (KCS) refers to any eye with degree of dryness.^[6]

The dry eye in SJS includes three important mechanisms: (1) aqueous tear deficiency, (2) decreased wettability of corneal surface, and (3) increased evaporation. Dry eye in SJS patients is often underestimated because; the punctum is closed due to

scarring or surgery. In SJS patients with severe dry eye, the dryness results in cosmic eye pain, and unstable tear film. Ocular pathologic process is often maintained by the destruction of goblet cells. Since the meibomian glands and goblet cells play a pivotal role in tear-film stabilization, this is likely to contribute to the evaporative effect of dry eye via the instability of tear film.^[7]

According to Ayurveda KCS can be corelate to Shushkakshipaka. One of the classifications given by Ācārya Suśruta for eye diseases is "Diseases affecting all parts of eyeball i.e., Sarvagata Netraroga; includes disease Shushkakshipaka - a very similar to Ocular surface disease i.e., Dry Eye Syndrome can be corelated to Shushkakshipaka described as a Vātaja disease in Sushruta Saṁhitā. [8] Vātapitta vitiated condition by Vāgbhaṭa [9] and Śārañgadhara considered this as Vātaraktaja. [10] In SJS mainly Vatapitta Prakopa along with vitiation of Rakta occurs due to Rooksha and Katu Ahara of patients. It will lead to evapouration of tear film and cause Toda, Beda and Upadeha to eyes and later eyelid became Rooksha and Daruna leads to difficulty to open and close eyes.

Corneal Ulcer and Perforation

A corneal ulcer, a defect of the corneal epithelium involving the underlying stroma, is a potentially vision-threatening ocular crisis.^[11] Even with instant treatment patients can suffer notable morbidity with complications including corneal scarring or perforation. There are two main components in the production of purulent corneal ulcer: Damage to corneal epithelium; and infection of the disintegrated area.^[12]

SJS due to infectious cause will directly lead to corneal ulcer. Corneal epithelial tight junction coherence depends on the interaction between cytokines derived from epithelial cells, stromal keratocytes, immune cells and lacrimal fluid and damage to tight junction coherence may agitate this equilibrium. [13] In SJS, corneal epithelial damage frequently occur which lead to corneal necrosis and later ulceration and perforation.

In SJS due to vitiation of Raktha and Pitta dosha in the body progressively cause disintegration of Krishna mandala and start damage in Drishti and Krishna.[14] If Krishna mandala (cornea) looks like that it is immersed in water (loses transparency or become edematous), pierced by needle (Corneal ulcer), profuse warm discharge from the eye with severe pain then it is called Savrana Shukla. By this elucidation it can be interpreted that basically there are four main symptoms described in Ayurveda for Savrana Shukla i.e., There is corneal edema. Ulceration of the cornea, profuse discharge which is warm in nature indicating acute inflammation of cornea. Severe pains which are normally found in intense infection of eve mainly associated with ciliary congestion due to rich sensory contribution of cornea.[15] In addition to Sushruta few more symptoms were described by Acharya Vagbhata in this disease, these are: Toda (Pricking sensation) Raga (conjunctival congestion), Pakwajambunibham (severely congested eye/ Ciliary congestion)_Prognosis of Savrana Shukla/Kshatta Shukla: in addition to the clinical features of the Savrana Shukla Aacharyas also described prognosis of Savrana Shukla based on location, types, number, features etc. Ulcers having following properties are considered as curable:[16] Ulceration away from central part of cornea viz. peripheral ulcer, Superficial ulcer, Absence of secondary infection, Painless Ulcer, Solitary ulcer. Ulcers having following properties are incurable:[17] Central corneal perforations, Corneal ulceration which are covered with granulation tissue, collateral existence of healed and newly formed ulcer are frequent presentation of protozoal corneal ulcer, Neovascularization of central cornea, Involving two layers i.e., involvement of vascular coat along with fibrous coat result into sequale and severe diminished vision. Intraocular haemorrhage /Neovascularization, having copious warm discharge with corneal abscess, Ulcer having grey, white color like wing of quail suggestive of severe necrosis of corneal layer. If ulcer in first layer i.e., Tejojalashrita Patala [18] of eyeball it is difficult to treat If it involves two layers i.e., Tejojalashrita and Mamsashrita Patala [18] then it is Yapya (if medication and proper care is taken it do not complicate). If it affects three layers [18] i.e., first two

coats followed by third fibrous tunic of eye which may be taken as Cortex of the lens.

Savrana Sukla can lead to Ajakajata (Perforated corneal ulcer)^[19,20] In this corneal disorder cornea gets penetrated with prolapsed iris which has given classical similarity with goat excrement. The clinical features include severe pain and profuse discharge. The prolapsed iris may be attributed classical black colour because of reduced extirpated blood and its contact with external ambience. According to Ayurvedic texts the prolapsed iris having distinct red colour, salty and thick blood mixed lacrimation with severe pain. It is Raktaja dominant incurable disease according to Sushruta while his treatise annotator Dalhana categorized it as Kaphaja predominant incurable disease^[21]

Superficial Punctate Keratitis (SPK)

Superficial punctate keratitis is characterised by occurrence of multiple, spotty lesions in the superficial layers of cornea^[22]

In SJS due to viral or bacterial infections can lead to ocular symptoms like keratoconjunctivitis, blepharoconjunctivitis which can cause SPK. Major source of mucin content is situated in goblet cells which help for the maintenance of tear film. In SJS destruction of goblet cells lead to keratoconjunctivitis sicca which later causes SPK. Meibomian gland dysfunction cause disruption of the tear film, causing severe dry eye and lead to SPK. [23]

Avrana Shukla means healed corneal ulcer, without opacity and the size of the opacity and ulcer is directly proportional. According to Acharya Sushruta a white spot appears in the cornea which is caused by Abhishyanda (Conjunctivitis) with painless condition, absence of lacrimation and like white cloud in the sky is Avrana Shukla. In context of prognosis, he has differentiated his opinion by saying that dense, indurated, and chronic in nature is said to be curable with difficulty. Avrana Sukla is Raktha Pradhana Sadhya Vyadhi. In SJS vitiation of Rakta dosha occurs due to various irregular habits and infectious cause leads to Abhishyanda of eyes and later Avrana shukla symptoms mainly white spots in Krishna Mandala. [24,25]

Corneal Keratinization

Keratinization of the tarsal conjunctiva in an eye with adequate tears occurs following several conditions, including irritation and susceptibility to topical medications, Stevens-Johnson syndrome, radiation to the lid, and occasionally from unknown causes. The keratinized cells produce an epithelial keratitis with subsequent vascularization of the cornea. [26]

SJS due to corneal epithelial defects, severe dry eye and keratitis can cause corneal keratinization.

In SJS later stages all the *Doshas* get vitiated vitiation gradually. Sira Shukla arises due to complication of corneal ulcer and described by *Vagbhata* only. This is found in chronic corneal ulcer. The disease is featured with corneal vascularization, pricking pain, burning sensation and copper coloured cornea assigned to vitiated *Rakta dosha*. There is serosanguinous discharge of assorted temperature of thick consistence without any cause. The disease at this stage shows little signs of recovery. The disease symptoms at this stage stays compatible with certain features of classical disorder termed as *Savrana Shukla*.^[27]

DISCUSSION

Steven-Johnson syndrome (SJS) is life-threatening hypersensitive mucocutaneous disease triggered generally by medication and infections. Even after recovering from skin problems, survivors can have serious ocular complications leading to blindness. Initially, eye care is often displaced by the lifethreatening dermatologic troubles. Nevertheless, the common and devastating long-term complications often are ophthalmic in character.^[28] As these complications can be reduced or even prevented with early intervention, an eye examination should be an essential component of the evaluation of any patient with suspected SJS. Despite this, patients may seek treatment from ophthalmologists many years after disease onset, and the dermatologic changes have resolved, and permanent visual impairment has occurred.

Most of the corneal pathologies in Stevens Johnson syndrome can be corelate to *Savrana* and *Avrana*

Shukla symptoms in ayurveda and Keratoconjunctivitis sicca can be correlate to *Shushkakshipaka* symptoms.

CONCLUSION

SJS is a dermatological emergency condition with a high incidence of ocular involvement. As observed corneal pathologies occurred most of the patients suffering from SJS. All the corneal defects are progressive stage of acute ocular manifestation of SJS. Chronic sequelae can be avoided by proper care and early interventions. SJS is a systemic disease, so Ayurveda line of treatment always manages the Samprapthi of disease and treats the corneal pathologies. Shushkashipaka mode of treatment can be adopted for KCS. Most of the corneal pathology symptoms can be managed by Avrana and Savrana shukla chikilsa. According to the disease symptoms prognosis is classified under Sadhya, Krichrasadhya, Yapya and Asadhya. In conclusion, all patients with SJS should undergo initial ophthalmologic screening during the acute phase of the disease to prevent the blindness due to cornea. The integrated disease approach, treatment application and predicting prognosis will satisfy the resultant outcome of the disease in a more favourable view which will augur a better perspective in managing preventable corneal blindness.

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