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Clinical Manifestations of Steven's Johnson Syndrome/Toxic Epidermal Necrolysis

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ABSTRACT

BACKGROUND: Toxic epidermal necrolysis (TEN) and Stevens-Johnson syndrome (SJS) both reflect extreme manifestations of the same clinical entity responsible for significant, life Threatening hypersensitivity mucocutaneous reactions, caused by intraepidermal cell death resulting in epidermal sloughing and blistering; often induced by medications. The mucous membranes of the mouth, stomach, lungs,skin, and genitourinary system are all major sites of Involvement. Despite receiving effective treatment at the local and systemic levels, some survivors may have major ocular consequences that might ultimately result in blindness. It is still unknown whether or not SJS/TEN can be effectively treated locally or systemically. If you want to improve your long-term eye health, it's crucial to catch any problems early and treat them aggressively. Chronic ocular surface failure, including a lack of limbal cells and full keratinization of the ocular surface, may be detected by inspecting the eyelid edge,palpebral conjunctiva, and fornix. It is possible to lessen the danger of ocular surface failure using transplants of amniotic membrane or a cultured oral mucosal graft.

AIM: The aim of this investigation is to learn more about the symptoms of SJS/TEN that emerge in the eyes.

METHODOLOGY: Patients with ocular signs of SJS/TEN who sought treatment at C.U.Shah medical college and hospital, a tertiary hospital, via the OPD and IPD of skin and VD were included. Between January 2022 and May 2022, all patients had a documented ophthalmologic evaluation.

RESULTS: The study had 17 participants. Ocular involvement was seen in 10 individuals, with 5 patients (50%) experiencing mild ocular indications, 2 patients (20%) experiencing moderate visual manifestations, and 3 patients (30%) experiencingsevere ocular signs. Two patients (20%) had involvement of the cornea, seven patients (70%) had involvement of the conjunctiva, and six

INTRODUCTION:

Extreme cutaneous adverse reactions include Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN), which occur at opposing ends of the spectrum. The affected region in mild SJS cases is less than 10% of the body's total surface area. TEN is the worst form since it covers so much of the body. There is a transitional group that consists of people who are both SJS and TEN participants (10-30%). Predisposing factors include fever and malaise, and the subsequent development of a broad, sensitive cutaneous eruption comprising macules, papules, unique target lesions, and vesicles or bullae. In the midst of severe dermatologic indications, a physician may fail to recognise the presence of ocular sequelae, which are permanent and destructive to visual acuity owing to damage of the ocular surface. Eye problems are common in patients with SJS and TEN; studies have revealed that anything from 50% to 88% of these instances entail acute ocular involvement. Acute ocular surface

Inflammation may have devastating long-term effects, including dry eye disease, recurrent or chronic corneal epithelial abnormalities, conjunctival scarring, symblepharon formation, a lack of corneal limbal stem cells, and corneal scarring. Concerns have been raised about the

consensus on the molecular pathophysiology of SJS/TEN. The drug's uniqueness means that various cultures and ethnic groups have distinct genetic risk factors. In addition, delayed drug hypersensitivity reactions to a medication or its metabolites seem to include cell-mediated apoptosis of keratinocyte via the FAS signalling cascade and granulysin synthesis in as many as 75% of instances. Infectious factors are probably to blame for the remaining 25%. The purpose of this research was to catalogue the ocular symptoms of SJS/TEN among tertiary care hospital outpatients and inpatients visiting the skin and VD OPD.

MATERIALS AND METHODS:

A purely descriptive research based on observation was carried out at C. U Shah medical college and hospital, Surendranagar and comprised data on age, gender, etiology, and ocular findings related to patients diagnosed with SJS/TEN between January 2022 and MAY 2022 was fetched. Patients were classified according to the criteria outlined: those with less than 30 percent BSA involvement were grouped together as SJS or SJS/TEN overlap, and those with 30 percent or more BSA involvement were categorised as TEN based on clinical history or skin biopsy findings. Records of ocular examinations were not included for patients who lacked them.

The research comprised 17 SJS/TEN patients who were seen at the C. U. Shah Medical College and Hospital's Department of Skin and VD for either outpatient or inpatient care. All patients gave informed permission before having their demographic information collected and undergoing a thorough dermatologic and systemic examination. Both the medicines suspected of causing the problem and the specifics of the treatment plan were documented. After the first evaluation, patients were followed up with on a regular basis for a total of 6 months.

In cooperation with the ophthalmology division, we screened all patients with an eye exam. Mild, moderate, and severe degrees of involvement at presentation were established using clinical criteria detected using an ocular slit-lamp examination. Slight ocular involvement included desquamation and denudation of eyelid skin, edoema of the eyelids, mucous discharge, mild corneal involvement, mild conjunctival injection. Mild instances included membranous conjunctivitis, epithelial defects that healed at a rate of less than 30% despite medical intervention, corneal infiltrates or ulceration. Eyelid malposition and symblepharon development were among the most severe symptoms, along with persistent corneal epithelial abnormalities, whole or partial vision loss, and a shortened conjunctival fomix.

RESULTS:

Nine men and eight women made up the total number of patients included in the research. Eleven had SJS or SJS/TEN overlap, while the remaining six had TEN. Ten individuals (or 90%) had ocular signs, with men outnumbering women by a ratio of 6 to 4. With the exception of a single instance (Table 1) where no evidence of drug use previous to the onset of lesions could be uncovered, all cases were caused by drugs. Every patient had symptoms in both eyes. Ocular symptoms such as swelling, watering, redness, buming, discharge, and sloughing of skin over lids were all present in the acute phase for all patients who reported to us. The above-described categorization method was used to assign grades of mild, moderate, and severe to the acute symptoms. Fifty percent (5 patients) were only mildly affected, twenty percent (2 patients) were moderately affected, and thirty percent (3 patients) were severely affected. Six patients (60%) had lid findings, seven patients (70%) had conjunctival findings, and two patients (20%) had corneal findings after a comprehensive ocular examination [Table 3]. Discharge, lid border ulceration and crusting, lid edoema, eyelash matting,

meibomitis, blepharitis, distichiasis, and lid peeling were some of the lid problems. Conjunctivitis, conjunctival membranes, subconjunctival haemorrhage, and symblepharon were all problems of the conjunctiva. Corneal ulceration, punctate epithelial keratitis, and punctate epithelial erosions were all types of corneal problems.

In the first, acute stage, all patients received topical corticosteroids, tear replacements and topical antibiotics.

Four individuals (29.6 percent) had persistent eye symptoms after the trial period ended. One of these patients suffered severe dry eye illness, two others experienced vision loss, and another experienced extreme photophobia. Ocular symptoms were modest to severe in patients with SJS and SJS-TEN overlap. It has been shown that individuals with early significant ocular involvement are more likely to have difficulties later on.

Ocular signs were more likely to occur in those who had suffered more extensive injury to their skin and oral mucosa.

DISCUSSION:

The purpose of this research was to examine the clinical manifestations of SJS/TEN, with a focus on ocular sequelae, at a large academic medical centre. Ocular morbidity is often brought on by SJS and TEN. Our study found that 58.82% of SJS/TEN patients had ocular abnormalities. The majority of our research participants had a drug-related aetiology, with phenytoin being the primary suspect.

The degree to which the eyes are affected by SJS/TEN may be categorised as mild, moderate, or severe. Desquamation and denudation of the eyelid skin, eyelid edoema, minor involvement of cornea, mucous discharge, mild conjunctival injection, and chemosis are all symptoms of mild ocular involvement. Membranous conjunctivitis, epithelial abnormalities that heal more than 30% with medical therapy, corneal infiltrates or ulceration characterise moderate instances. Eyelid malposition may develop over time, symblepharon can occur, corneal epithelial abnormalities don't repair, vision loss might be total or partial, and the conjunctival fornix can shrink. About half of our patients came with just minimal ocular involvement, whereas a quarter had moderate involvement and a third had severe involvement

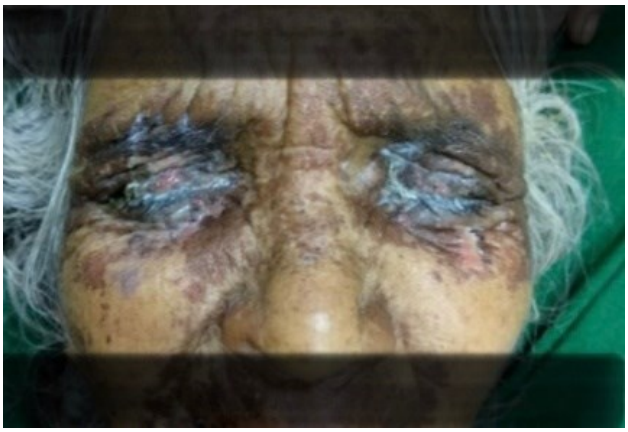
The clinical phases may also be used to categorise the ocular characteristics into acute, subacute, and chronic categories. Conjunctivitis, meibomitis, haemorrhage in conjuctiva, conjunctival membrane development (or false membrane development), symblepharon, and abnormalities of epithilium are all part of the acute stage, which typically lasts for up to 2 weeks following the beginning of symptoms. Chronic cicatrizing conjunctivitis, trichiasis, or distichiasis, as well as defects of corneal epithelium, stromal scarring, and infection may result from infiltration and inflammation of the ocular surface that does not resolve after the acute phase. Subacute conditions include these. Up to 30 percent of those with SJS/TEN have persistent ocular abnormalities. The most prevalent sites of involvement are the meibomian glands and the palisades of Vogt found inside the limbus. Keratinization of the bulbar and tarsal conjunctival surfaces, punctual occlusion, eyelid edge and persistent symblepharon and ankyloblepharon are further examples of chronic ocular sequelae. The current investigation found that all 10 patients (58.82%) came to us during the acute phase of their condition, and that 30% of these patients went on to acquire chronic sequelae by the conclusion of the study period. Eighty-one out of 117 individuals with acute ocular involvement were found in a research by Yip et al (69 percent). Fifty percent of the 44 participants those who were tracked for more than 6 months, had late problems.

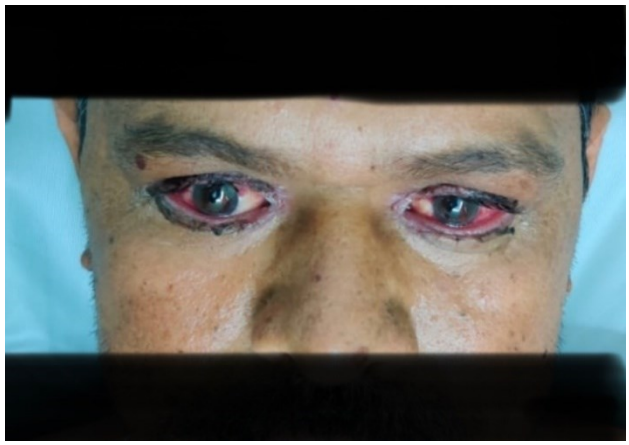
Prevention of the ocular symptoms and it's recovery depend on early ophthalmic examination and care as well as regular follow-up. Topical cyclosporine, topical broad-spectrum antibiotics, and topical corticosteroids are often used along with preservative-free lubricants to treat acute SJS/TEN. It has been suggested that using topical steroids early on might lead to better eye health and vision. Amniotic membrane transplantation is an effective early surgical technique

that has been shown to considerably improve epithelialization, minimise inflammation, and lessen scarring on the ocular surface in situations with more severe involvement. The primary goals of therapy during the chronic phase are the prevention of permanent damage to the ocular surface, the management of ocular sequelae, and the restoration of visual function. The loss of vision caused by structural problems may be corrected with surgical procedures including keratoprosthetics and keratolimbal allografting (KLAL). Cultured oral mucosal epithelial transplantation (COMET) and Limbal stem cell transplantation (LSCT) are advised for severe instances of dry eye and corneal blindness at the end stage.

CONCLUSION:

SJS and toxic epidermal necrolysis (TEN) despite their varying degrees of severity, are treated as a single illness. Common triggers for SJS/TEN include pharmaceuticals, infectious agents, and maybe some other unknown reasons. Although the exact cause of SJS/TEN remains unknown, some genetic predispositions have been found that differ across ethnic groups and between the medicines known to cause the disease. Our research showed that ocular symptoms occurred in 58.82% of SJS/TEN patients. Total 73.85% of patients had an acute ocular symptom, most often conjunctivitis, whereas thirty percent experienced chronic sequelae. Since no medication has been found to date that can stop the course of skin detachment, supportive care is likely more important than particular immunomodulatory therapies in improving the patient's condition. The severity of the illness, the age of the patients, and the presence of other medical disorders all contribute to the high death rate that persists despite treatment attempts. Long-term complications, such as mucous membrane strictures, may cause serious eye difficulties in survivors. Therefore, it is essential that patients with SJS/TEN get treatment and follow-up from a team of professionals from several disciplines.





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