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## Acute and severe ocular manifestations of a patient with Stevens-Johnson Syndrome in Wangaya General Hospital: a case report



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### ABSTRACT

**Background:** Stevens-Johnson syndrome is a severe mucocutaneous disorder with high mortality rates and is potentially life-threatening. Ocular manifestations in SJS occurred up to 81%, ranging from conjunctiva hyperemia, cicatrizing conjunctivitis with symblepharon, corneal ulceration, corneal blisters and perforation, which can potentially lead to permanent vision loss. This case study aims to evaluate the acute and severe ocular manifestations of a patient with Stevens-Johnson Syndrome in Wangaya General Hospital.

**Case Description:** The ophthalmologist department consulted a 34-year-old male patient with the diagnosis of Stevens-Johnson syndrome. He complained of bilateral upper and lower eyelid palpebral conjunctiva adhesion to bulbar conjunctiva. The

ophthalmological examination could not be examined due to the adhesion. The patient performed bilateral symblepharon separation of the eyelid using blunt-tip of surgical scissors under local anesthesia. After the procedure, the visual acuity was around 3/60 for both eyes. The anterior segment examination in both eyes showed that the palpebral was edema, peeling of the skin. The conjunctiva appears to be chemosis, conjunctival injection, with a mucopurulent secret in both eyes. The patient was treated with antibiotics, steroid eye ointment, and eye drops.

**Conclusion:** Ocular manifestations of SJS are mostly mild-moderate. However, several cases possess grave complications and may progress to vision loss. Treatment for ocular sequelae can reduce the risk of long-term visual morbidity.

**Keywords:** Stevens-Johnson Syndrome, Ocular manifestations, Antibiotics, Management.

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### INTRODUCTION

Stevens-Johnson syndrome (SJS) is a rare and life-threatening condition. The incidence in the United States has been reported at around 2.6 to 6.1 cases per million people annually.<sup>1</sup> In Indonesia, the incidence rate is unknown. A single center reported 75 cases over 15 years period.<sup>2</sup> It is a severe hypersensitive reaction with high mortality rates and is potentially life-threatening.<sup>1</sup> The mortality rate was 9.5% globally, but it can progress to Toxic Epidermal Necrolysis (TEN) with a mortality rate of 13.6%.<sup>1,2</sup> Patients with previous SJS/TEN reactions suffer high-risk, long-term complications, including

skin, eye, mucosal, respiratory, renal, and/or hepatic systems.<sup>3</sup>

SJS is an immunological event that is classified to type IV hypersensitivity. It is a severe mucocutaneous disorder that can severely affect the skin, the oral, the ocular, the genital, the gastrointestinal, and the anal.<sup>1,4,5</sup> Drugs are among the most common etiology of SJS, besides a bacterial infection, vaccination, or graft-versus-host disease.<sup>6,7</sup> Medications that most commonly cause SJS are certain antibiotics, acetaminophen, anticonvulsant drugs, allopurinol, sulfonamide drugs, non-steroidal anti-inflammatory drugs (NSAIDs), and other drugs.<sup>7-9</sup> Most of these drugs are commonly used, but no

one can predict which drugs will cause a severe adverse reaction like SJS.<sup>6</sup>

Clinical manifestation in SJS usually begins with influenza-like symptoms such as malaise and fever, followed by severe mucocutaneous lesions. Around 80% of cases involve mucous membranes, including oral, ocular, gastrointestinal, genital, and anal.<sup>8</sup> Ocular manifestations in SJS occurred up to 81%, ranging from conjunctiva hyperemia, cicatrizing conjunctivitis with symblepharon, corneal ulceration, corneal blisters, and perforation, which can potentially lead to permanent vision loss.<sup>8</sup> Bilateral conjunctival hyperemia with purulent discharge is the most common ocular

finding in the acute phase.<sup>9,10</sup> Development of symblepharon can be found in the sub-acute and chronic phases of SJS. It can be partial or complete symblepharon adhesion from palpebra conjunctiva to bulbar conjunctiva. In some conditions, symblepharon can obstruct the lacrimal duct and interfere with the tear flow, causing severe dry eye.<sup>4,11</sup> Furthermore, these ocular complications commonly cause chronic ocular sequelae, which lead to long-term pain and discomfort for the patient.<sup>11</sup> Hence, the authors want to report this case weighing the importance of rapid diagnosis and initial treatment to save the vision.

## CASE REPORT

A 34-year-old male patient was being consulted to the ophthalmology department with a complaint of bilateral upper and lower eyelid palpebral conjunctiva adhesion to bulbar conjunctiva. He was admitted to the emergency department (ER) one day prior with chief complaints of a burning sensation all over his body, including both eyes, oral ulcers, and macular rash on his chest toward the extremity after taking oral amoxicillin and acetaminophen. The patient denied headaches, nausea, and vomiting. He was hospitalized in the dermatologist ward with Stevens-Johnson Syndrome (SJS) diagnosis. On admission, the patient was febrile with a body temperature of 38°C and tachycardic with a pulse of 118 beats per minute. Other vital signs were within normal limits, with a blood pressure of 125/78 mmHg, respiratory rate of 20 breaths per minute, and oxygen saturation of 97% room air.

Initially, ophthalmological evaluation was unable to be performed due to severe eyelid adhesion. Therefore, the patient was diagnosed with symblepharon Oculi Dextra et Sinistra (ODS) and was performed bilateral symblepharon separation of the eyelid using a blunt tip of surgical scissors under local anesthesia. After the procedure, the symblepharon was completely removed, and an ophthalmological examination was performed. His uncorrected visual acuity was around 3/60 for both eyes. The anterior segment examination in both eyes showed that the palpebral was edema,



**Figure 1.** Acute ocular manifestation showed bilateral upper and lower eyelid palpebral conjunctiva adhesion to bulbar conjunctiva, palpebral edema, crustae, and skin peeling.



**Figure 2.** Acute ocular manifestation showed palpebral edema, crustae, sloughing of skin, inflammation of both eyelids, conjunctival injection, chemosis, mucopurulent secret in both eyes

crustae, and skin sloughing. There is an erythematous and edema of the eyelid due to inflammation. The conjunctiva appears to be chemosis, conjunctival injection, with a mucopurulent secret in both eyes. At the same time, the cornea and other anterior segments appear to be normal. The posterior segment in both eyes could not be visualized.

The patient was then diagnosed with symblepharon and blepharoconjunctivitis of both eyes due to SJS (Figures 1 and Figure 2). For initial treatment, a combination of Dexamethasone, Neomycin Sulfate, Polymyxin B Sulfate eye ointment twice daily, and Tobramycin eye drops every 3 hours for ocular management. The patient and his family were also educated to keep his eyes hygienic. The patient was also planned for the next follow-up a week post-discharge.

## DISCUSSION

SJS is a severe dermatological condition and a medical emergency with a high

mortality rate involving skin and mucous membranes. The study by Abrol et al. showed that up to 90% of the patients suffering from SJS had ocular involvement. An ocular manifestation of SJS is classified as mild, moderate, and severe. Mild manifestation is indicated by eyelid skin involvement in the form of desquamation and denudation, palpebral edema, mild corneal involvement, injection of the conjunctiva, mucous secret, and chemosis. Membranous conjunctivitis, epithelial defects with more than 30% healing with medical treatment, and corneal ulceration are included in the moderate stage. Severe manifestations of ocular SJS include eyelid malposition, formation of symblepharon, nonhealing corneal epithelial defects, complete or partial vision loss, or foreshortening of conjunctival fornix.<sup>12</sup>

According to clinical stages, ocular SJS is classified into acute, subacute, and chronic. The acute stage occurred 2 weeks after the initial SJS symptoms. The acute stage comprises conjunctivitis

or conjunctival hemorrhage, formation of conjunctival membrane pseudomembrane, meibomianitis, symblepharon, and epithelial defects, which then resolves over time.<sup>12</sup> In some cases, despite resolution after the acute phase, persistent inflammation and ulceration of the ocular surface may lead to the subacute stage, indicated by chronic cicatrizing conjunctivitis, trichiasis, or distichiasis, even corneal epithelial defects such as infection, stromal scars. Chronic ocular changes occurred in up to 35% of SJS/TEN cases with the involvement of palisades of Vogt within the limbus and the meibomian glands. The other less common chronic ocular sequelae include permanent symblepharon and ankyloblepharon, cicatricial entropion, punctal occlusion, keratinization of the eyelid margin, tarsal, and bulbar conjunctival surfaces.<sup>12</sup>

In this case, the patient was found to have severe manifestations in the acute stage, such as the formation of symblepharon followed by other ocular manifestations such as palpebral edema, crustae, sloughing of skin, erythematous and edema of both upper and lower eyelid and the conjunctiva showed chemosis, injection, and mucopurulent secret in both eyes. The absence of complaints of headache, nausea, and vomiting indicates no symptomatic glaucoma complications. We should be aware of glaucoma, uveitis, and infectious endophthalmitis since it is one of the complications in the chronic phase of SJS.<sup>12</sup>

Drugs are the most common causative agent for SJS, and antibiotics are the most common cause. A study by Yang SC et al. found that the Penicillins group is the most common causative antibiotic of SJS in China (over 7%), similar to Singapore (almost 12%) and Thailand (over 30%). However, Sulfonamide is the most common group of antibiotics responsible for SJS reaction in Malaysia (over 17%) and the Philippines (over 7%).<sup>1</sup> Study by Fakoya AOJ et al. found that Penicillins are the most frequent causes of SJS compared to the other antibiotics. The variation may be due to the frequency of Antibiotics being prescribed in a certain region.<sup>13</sup> Before SJS, the patient had a history of taking Amoxicillin and Acetaminophen without

a prescription. Amoxicillin is most likely responsible for the given allergic reaction because, from the history, the patient had never taken Amoxicillin. In contrast, the patient had used acetaminophen before and was given it during hospitalization without any allergic reaction.

Early recognition of symptoms, discontinuation of all suspected culprit drugs, and prompt treatment, followed by persistent follow-up, are the main factors for reducing mortality and improving prognosis, as also recovery and prevention of the ocular manifestations.<sup>11,12</sup> Using systemic corticosteroids as an SJS therapy is still controversial. Several studies have argued that corticosteroids will reduce the severity of the disease and prevent severe complications, disease progression, and recurrence. Some literature states that systemic corticosteroid administration can reduce inflammation by improving capillary integrity, promoting the synthesis of lipocortin, and suppressing the expression of adhesion molecules. In addition, corticosteroids can regulate the immune response through down-regulation of cytokine gene expression. A study by Grunwald P et al. showed that short-term corticosteroids might benefit survival rates.<sup>14</sup> A study by Kim et al. showed that a high-dose systemic steroid therapy used during the acute phase could improve visual outcomes in the long term.<sup>10,14</sup> A prospective study to evaluate the impact of systemic intravenous steroids combined with topical steroids showed that if half of the patients had severe ocular involvement with corneal ulceration, after 1 year, they had no ocular sequelae and relatively good visual acuity. Sotozono C et al. observed five cases with ocular manifestation in SJS treated with systemic and topical steroids within four days from disease onset. They showed that none of the cases had visual dysfunction.<sup>10,15</sup>

On the other hand, several authors do not agree with using corticosteroids as an SJS treatment, arguing that corticosteroids will inhibit wound healing, increase the risk of infection, mask early signs of sepsis and gastrointestinal bleeding, and increase mortality. Another factor that must be considered is tapering off corticosteroids.<sup>11</sup> Although infection prophylaxis still lacks treatment evidence,

topical antibiotics may be needed to treat brief bouts of inflammation.<sup>1</sup> Furthermore, research showed that if there is no sign of improvement in 3-5 days, corticosteroids should be discontinued.<sup>11</sup>

The treatment in acute ocular SJS combines topical corticosteroids, topical Cyclosporine, and broad-spectrum topical antibiotics with the concurrent use of preservative-free lubricants. Early use of topical corticosteroids may be associated with improved visual outcomes.<sup>12</sup> Early ocular treatment is crucial in preventing an ocular immune response responsible for long-term scarring.<sup>14</sup> Wound toilet should be performed daily, using a cotton-tip applicator and saline flush to prevent conjunctiva adhesion of the eyelids.<sup>16</sup> In addition to topical antibiotics, corneal epithelial defects may be prevented with lubricant to aid healing and minimize trauma.<sup>16,14</sup> In this case, the patient was given a combination of Dexametason, Neomycin Sulfate, Polymyxin B Sulfate eye ointment, and Tobramycin eye drops.

Severe dry eye disease, diminution of vision, and trichiasis are among the most commonly found long-term ocular sequelae after 6 months. The main strategy for the chronic phase is mainly about preventing further ocular surface damage, managing the ocular sequelae, and possibly visual rehabilitation. Several structural abnormalities, such as keratoprosthesis and Keratolimbal Allograft (KLAL), sometimes require surgical interventions to allow visual recovery. Severe cases which poor prognosis, such as corneal blindness and severe dry eye, may require limbal stem cell transplantation (LSCT) and cultivated oral mucosal epithelial transplantation (COMET).<sup>12</sup>

This case report discussed a case of ocular manifestation of Stevens-Johnson Syndrome, which is handled at the secondary level hospital. The limitation of this case is the unavailability of standard objective visual acuity examination and further ophthalmic examination, which can only be performed in the clinic.

## CONCLUSION

An ocular manifestation of SJS is mostly mild-moderate. However, several cases possess grave complications and may progress to vision loss. Treatment for



these ocular sequelae can reduce the risk of long-term visual morbidity. Knowing the etiology and progression of the disease is necessary for early diagnosis and prompt treatment. Self-awareness about consuming drugs only when medically indicated is crucial in preventing side effects, including severe immune reactions.

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## CONFLICT OF INTEREST

The author stated there is no conflict of interest in this study.

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## AUTHOR CONTRIBUTION

All of the authors equally contributed to the study.

## ETHICAL CLEARANCE

Patient approval has been obtained in this study.

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