

CASE REPORT

CASE REPORT: BLEPHAROCONJUNCTIVITIS AS MANIFESTATION OF STEVENS-JOHNSON SYNDROME

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ABSTRACT

Background: Stevens Johnson syndrome (SJS) is a collection of acute and life threatening skin symptoms caused by an allergic drug reaction. SJS also attacks the other mucous membranes, one of which is the eye. SJS is a rare case. The initial complaint of SJS is the eruption on the skin in the form of redness followed by blisters and attacking other mucous membranes. The ocular manifestations of SJS are conjunctivitis, corneal erosion, corneal ulcers, and blepharitis.

Methods: Treatment of SJS's ocular manifestation is administration of drugs and surgical intervention. Administration of drugs that often done in the acute phase is antibiotic eye drops with steroids to prevent complications. Complications that can occur include symblepharon, distichiasis, and keratitis. This complication can cause a sharp decrease in vision.

Results: This case report discusses a 68 year old male patient with blisters all over his body accompanied by swelling in both eyes, redness in both eyes, and difficult to open eyes. Patients with history of taking the new drug a month ago. No history of drug allergy. The patient suffered from hypertension, non-hemorrhagic stroke, and epilepsy with irregular treatment. Patient was performed a fluorescein test. In this case, the patient is given antibiotic ointment and artificial tears with steroids. In this case there were no complications.

Conclusion: The ocular manifestations of SJS if handled quickly and properly can prevent long-term complications.

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Introduction

Stevens Johnson Syndrome (SJS) is life threatening condition and rarely happened. Incidence of SJS in Western countries 0,1-1 in 100.000 population¹. SJS in Indonesia happened at least 12 cases on a year. Mortality rate for SJS can reached 5-15% in total cases². In other study, SJS mortality rate usually on elderly patient. Pediatric patient have a lower mortality rate than geriatric patient, but pediatric patient more likely experienced long term complication³. Patient with SJS tend to have ocular manifestation and the acute phase of ocular manifestation occur in 50-88% cases. The acute sign of ocular involvement which result from long term sequelae are cornea and conjunctiva epithelial defect, dry eye, symblepharon, cornea scar, and cornea limbal stem cell deficiency⁴. The authors are interested in discussing SJS case because of frequently seen ocular cases in SJS.

Case

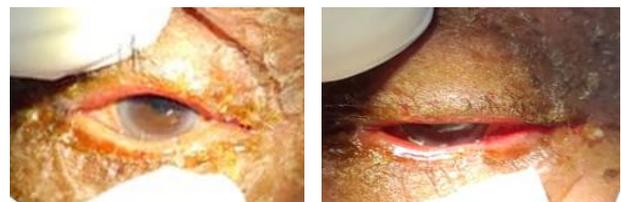
A 68 years old male, married, Balinese, Hindu, had been work as a teacher was referral patient from the Aricanti Hospital, Ubud, Gianyar, Bali.

Medical History

The patient came complaining of full body blisters since 3 days ago. Initially, the patient had fever for 4 days ago. The patient also complained of reddening of the whole body and received therapy in the form of azithromycin, paracetamol and vitamins since 3 days ago. After that, the patient complained of blisters all over his body. The whole body is reddish and turns white scaly and sloughing. Other complaints include swelling and redness of the patient's eyes. Redness of eye

appeared when the body full of blisters. It began when the patient complained of swelling in both eyes followed by redness of the eyes. The patient's eyes were also difficult to open and accompanied by pain. The patient also complained of white eye discharge, sometimes yellowish white eye discharge. Patient was not complaining blurred vision, photophobia, and watering. Patients also had hypertension, epilepsy, and non-hemorrhagic stroke with treatment of atorvastatin, amlodipine, and phenytoin. This disease has been suffered since 2019. The patient rarely used his medicine and began to reuse the drugs on January 6, 2021. There is no history of wearing glasses or eye disease on this patient. Previously, the patient had no drug allergy. The patient's family history is the patient's parents suffering from parkinson's disease, hypertension, and stroke. The patient's sister was suffering from hypertension. Ocular family history was not found.

Figure 1. The patient's eye condition on the first



day of hospitalization. Crusted on the right and left eyelids. The right and left eyes looked difficult to open.

Physical Examination

On physical examination, found uncorrected visual acuity for both eyes are $>3/60$. On bed side external examination, found swollen of superior and inferior palpebra with crusting of the eyelid, sloughing of skin, and erythematous macules. On examination of the anterior segment of the eye, it was found conjunctiva vascular injection. The

conjunctiva palpebra was difficult to evaluate. The cornea is clear. The anterior chamber, iris and lens are normal. The patient also underwent an additional examination of a fluorescein test and the result was there were no defects in the cornea and conjunctiva epithelium. Patient diagnosed with blepharoconjunctivitis.

Treatment

Blepharoconjunctivitis was treated with eye drops containing dextran-70 1 mg and hypromellose 3 mg given 6 times a day to both eyes and eye ointment containing gentamycin which was given 2 times a day to both eyes.

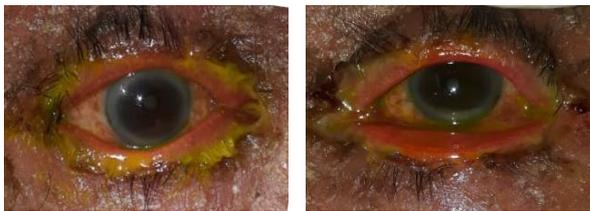


Figure 2. Fluorescein test negative

Follow Up and Outcomes

In the patient's daily follow-up, the patient showing improvement of visual problems from day to day. The patient said complaints of swelling and redness in both eyes improved. Then, the complaints of eye discharge are said to have started to decrease. Complaints of pain in both eyes were also said to have decreased. This patient did not experience complications such as defects in the cornea and conjunctiva, and a sharp decrease in vision. There are no adverse and unanticipated events that have not been handled properly in this case. The prognosis in this patient is *dubia ad bonam*. The patient and the patient's family already understand about the disease and how to prevent complications from this disease. The patient's and patient's family really maintains eye cleanliness and is assiduous in using the drugs

used, gentamycin eye cream and dextran-70 1 mg and hypromellose 3 mg eye drop.

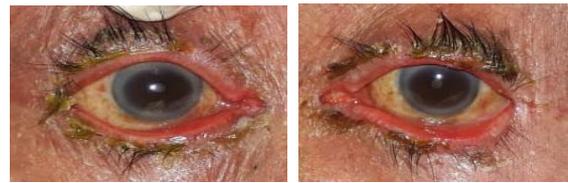


Figure 3. The patient's eyes on the third day of treatment. The eyelids did not appear swollen and the redness in both eyes improved.

Discussion

SJS is an acute and life-threatening skin drug reaction that affects the skin and other mucous membranes⁵. SJS is more common in young adults, but can affect other age groups. Acute symptoms of SJS in the eye include redness of the eye, sensation to having a particle of sand in the eye, photophobia, watering, and blurring⁶. Jenkins's study explained eye symptoms in the acute stage often occur with 15% -75% bilateral conjunctivitis and 25% conjunctiva or corneal ulceration⁵. This case is a 68 year old man with complaints of swelling in both eyes since 3 days ago. This complaint began with the blister on whole body of the patient after experiencing fever for 4 days. Swelling in both eyes accompanied by redness of the eyes. The patient also has white discharge in the eye. The patient also said it was difficult to open both eyes and felt pain. The patient did not complain of blurring of the eyes, photophobia, and watering. Several types of drugs that can cause Stevens Johnson Syndrome, namely antibiotics such as sulfonamides and trimethoprim, analgesics such as paracetamol and NSAIDs, HIV treatment with combination therapy using nevirapine, barbituates, allopurinol,

and anticonvulsants such as carbamazepine, phenytoin, and lamotrigine^{5,6,7}.

In this case, patients took medication of azithromycin, atorvastatin, amlodipine, and phenytoin. The patient started taking medication again during the past month due to patient non-compliance in taking regular medications such as atorvastatin and amlodipine and vitamins. The drug he recently drank in the form of phenytoin was due to complaints of seizures that he had just suffered in the past month.

Based on other literature, symptoms of SJS in the eye are frequent and can cause visual impairment, so patients with SJS should be evaluated immediately⁷. Initial symptoms of SJS occurred in the prodromal phase, namely malaise and fever followed by soft skin eruptions and generally consisted of macules, papules, vesicles, and bullae. Another symptom is that at least two other mucosal surfaces are affected such as erosions and crusting of the mouth, conjunctivitis, and urogenital manifestations such as urinary retention and urethritis which may accompany skin manifestations in 90% of cases⁸. On a slit-lamp examination, the severity of eye manifestations in SJS can be determined as mild, moderate, and severe^{8,9}. Mild eye manifestations consist of desquamated and bald skin of the eyelids, edema of the eyelids, mild conjunctival injection, mucus discharge or chemosis. Moderate eye manifestations may include membranous conjunctivitis, epithelial defects with more than 30%, corneal ulceration, or corneal infiltrates. Severe ocular manifestations consist of acquired eyelid malposition, symblepharon, non-healing corneal epithelial defect, partial or complete loss of vision, or

narrowing of the conjunctival fornix. Chronic ocular sequelae due to SJS occurs in 21-29% of pediatric cases and 27-59% of adult cases⁸.

Additional investigations such as fluorescein stain should be performed to evaluate for corneal and conjunctival epithelial defects⁷. Patients have symptoms such as persistent ulceration, dry eyes, and scarring of the eye as much as 35% due to chronic eye damage⁵. Patient in this case was in the acute phase. Patient was carried out several physical examinations as well as a supporting examination in the form of a fluorescein test on the patient. The uncorrected visual acuity for both eyes are $>3/60$. On bed side external examination, found swollen of superior and inferior palpebra with crusting of the eyelid, sloughing of skin, and erythematous macules. On examination of the anterior segment of the eye, it was found conjunctiva vascular injection. The conjunctiva palpebra was difficult to evaluate. The cornea is clear. The anterior chamber, iris and lens are normal. The patient also underwent an additional examination of a fluorescein test and the result was there were no defects in the cornea and conjunctiva epithelium.

Ocular treatment in SJS is divided into 2. There are drugs and surgical management. Management of Stevens-Johnson's acute ocular manifestations is administration of topical antibiotics, steroids, calcineurin inhibitors, and lubricants⁸. Use of topical steroids at the onset of symptoms can result in better eye visuals. In severe cases, early surgical intervention can involve transplanting the amniotic membrane, which can increase epithelialization and reduce inflammation and scarring on the surface of the eye¹⁰. One of the amniotic membrane transplant methods is called

cryopreserved amniotic membrane, which is a biological device made by cutting the amniotic membrane between two symblepharon rings and a human amniotic membrane graft. There have been several studies conducted to evaluate amniotic membrane transplantation in the eye in SJS and it appears to be showing significant results. The cryopreserved amniotic membrane produces maximum results when applied early in the disease by reducing the risk of long-term eye damage and producing sequelae after 6 days⁵. On the other study, treatment for the chronic phase aims to prevent worse eye surface damage, treat eye sequelae, and visual rehabilitation. If there are structural abnormalities of the eye, surgical interventions such as keratoprosthesis and keratolimbus allografting (KLAL) are required to help restore the visual eye. In the final stages with severe corneal blindness and dry eye, limbal stem cell transplantation (LSCT) and cultivated oral mucosal epithelial transplantation (COMET) are recommended. Providing prompt diagnosis and treatment is the key to good eye prevention and recovery in SJS cases^{8,10}.

On the Pandiaraj case series, eye management with SJS was carried out with a frequent topical lubricant, carboxymethyl cellulose, in all patients. Then, given a topical antibiotic in the form of tobramycin if secondary infection is suspected. Fluorometholone can be added for patients with excessive eye inflammation. In patients with symblepharon, membranolysis with glass rod passing is performed 2-3 times a day. All patients were asked to move their eyes and frequently asked to separate their eyelids to prevent symblepharon. Eye hygiene must be maintained properly. Patients with corneal epithelial defects

are given bandaged contact lenses. All patients were given systemic steroids while paying attention to the administration of fluids and electrolytes according to the patient's condition. This therapy is done once or twice a day for each patient until the patient goes home. No deaths were found and all were discharged within 2 weeks to 2 months¹⁰. In present case, patient was treated with eye drops containing dextran-70 1 mg and hypromellose 3 mg given 6 times a day to both eyes and eye ointment containing gentamycin which was given 2 times a day to both eyes. Currently, there are no complications in this case. Wang's study explained among those who survive, there are long-term eye complications that can become serious, which occur in about 60% of patients. Corneal and conjunctival damage in the form of scar tissue can lead to further visual disturbances⁷.

In Abrol's study, several complications were found in the form of complications on the eyelids, conjunctiva, and cornea. Complications on the eyelids include eyelid edema, discharge of eye discharge, distichiasis, ulceration of the edge of the eyelids and crusting, wrinkled eyelashes, meibomitis, blepharitis, and peeling of the skin above the eyelids. Conjunctiva complications include subconjunctiva bleeding, conjunctivitis, and symblepharon. Corneal complications include superficial epithelial keratitis, corneal ulceration, and erosion of the epithelium⁸. In the present study patient's daily follow-up, the patient showing improvement of visual problems from day to day. This patient did not experience complications such as defects in the cornea and conjunctiva, and a sharp decrease in vision. There are no adverse and unanticipated events that have

not been handled properly in this case. The prognosis in this patient is *dubia ad bonam*. The Abrol's study also found 8 patients who had chronic sequelae from the eye manifestations of SJS. Of the 8 patients, 3 patients had severe dry eye disease, 2 patients had trichiasis, 2 patients had decreased visual acuity, and 1 patient had severe photophobia. Complications are often not seen at first in conditions of severe eye manifestations. Damage to the mucous membranes of the skin and mouth was more associated with the incidence of ocular manifestations in SJS^{8,10}.

This case report discussed a case of Stevens Johnson Syndrome which is handled at the district level hospital and can be used as a reference for other research on eye cases in Stevens Johnson's syndrome and informs the hospital about things that need to be improved in handling rare cases like this. The limitation of this case report are that it does not include follow-up of patients after being discharged from the hospital and does not use snellen charts for visual examination due to limited facilities and infrastructure.

Conclusion

We discussed eye involvement in the case of Stevens Johnson syndrome with a 68 year old male patient who complained of swelling of the eyelids and redness of both eyes with blister all over his body after taking the new drug he had just taken for the past 1 month. The patient was also investigated with a fluorescein test and the result was that there were no defects in the cornea and conjunctiva. The patient was given antibiotic eye ointment and artificial tears with steroids to reduce inflammation in both eyes. The patient did

not experience complications, namely a sharp decrease in vision and defects in the cornea and conjunctiva and the formation of symblepharon. From this case, the author is more aware of a disease that is rare in cases but can be life threatening condition and produce long-term complications if not treated quickly and appropriately.

Conflict of Interest

The author stated there is no conflict of interest

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