CASE REPORT

SCLERAL CONTACT LENS FOR STEVEN-JOHNSON SYNDROME: A CASE REPORT

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ABSTRACT

Background: *Steven-Johnson syndrome is one of life-threatening skin abnormality. SJS complication involve multiorgan disfunction such as mucosa, ocular, respiration, and digestive tract. Around 30-50% of SJS patient experience chronic ocular sequel, which concludes progressive symblepharon, margo palpebral keratinization, trichiasis, entropion, corneal pannus, dry eye syndrome, and persistent epithelial cornea defect. Scleral contact lenses has beneficial in managing SJS patient, on both acute and chronic phases.*

Methods: *A 36-year-old female patient with SJS, presented for eye examination due to unpleasant sensation on both of their eyes and unsatisfactory quality of her vision. She presented high myopia, margo palpebral keratinization with trichiasis and distichiasis, symblepharon and hyperemic conjunctiva, wide corneal scar, and ocular neovascularization. A diagnostic trial set was used in the fitting process, and she was assessed according to standardized fitting methodology. Visual acuity, corneal topography, biometry, and ocular aberrations were evaluated. The follow-up period was 12 months.*

Results: *The best corrected visual acuity was 20/60 with correction lens S – 1.00 D for the left eye (LE) and 20/25 cannot be corrected with correction lens for the right eye (RE). After corneal contact lens fitting, visual acuity was improved to 20/20 and 20/25 for the RE and LE, respectively. The patient wore these contact lenses an average 8 hours a day. The patient's dry eye complaint was significantly decrease with more comfortable ocular sensation*

Conclusion: *The present case report describes how the patient had scleral contact lens fitted successfully for management of SJS ocular complication. They provided optimal visual quality and restore the patient's comfort.*

Keywords*: Scleral Contact Lens, Steven Johnson Syndrome, Dry Eye*

INTRODUCTION

Steven-Johnson syndrome (SJS) known as life-threatening skin abnormality, which contain of blister formation, skin loss and multi organ disfunction—generally, due to pharmacological treatment.[1] Steven-Johnson syndrome is the lightest clinical spectrum of acute mucocutaneous reaction, with skin rash as an early symptom which mimicking viral infection, then it develops into skin eruption in few days and the mucosa would be worsen. The incidence of SJS is rarely found, merely 1-6 cases per 1 million population every year.[2]

Supportive treatment of SJS focused on protecting skin from irritation and infection, pain reliever, preserve electrolyte and nutrition management. SJS' complication not only involves skin dysfunction, but also involve multiorgan dysfunction such as mucosa, including ocular surface.[1] Around 30-50% of SJS patient experiences chronic ocular sequel, which concludes progressive symblepharon, eyelid margin keratinization, trichiasis, entropion, corneal pannus, dry eye syndrome and persistent epithelial corneal defect.[3,4]

Scleral contact lenses known as large-diameter lenses that completely enclose the cornea, shielding it with a fluid reservoir to provide the perfect ocular surface environment, and perfectly useful for ocular surface diseases. According to the Scleral Lens in Current Ophthalmic Practice Evaluation (SCOPE) study group, ocular surface disease, corneal irregularity, and uncomplicated refractive error account for 16%, 74%, and 10%, respectively, of scleral lens prescriptions.[5] A prior study shown the therapeutic benefits of these lenses in the treatment of ocular surface diseases.[6] Scleral contact lenses can also be used to manage SJS patient, on both acute and chronic phases. The purpose of using scleral contact lens during chronic phase is to prevent corneal damage that caused by palpebral margin keratinization.[7-9] Previous studies revealed that scleral contact lens usage on patient with history of SJS can increase their visual acuity.[7]

This paper presents a case of patient with history of chronic SJS who develops severe dry eyes complaint, symblepharon and eyelid margin keratinization which causes wide corneal scar. Scleral contact lens was given to the patient, in terms to manage the patient's dry eye complaint and improve the visual acuity.

CASE DESCRIPTION

A 36-year-old female patient was admitted to eye hospital with chief complaint of ocular pain on both of her eyes, dazzled eyesight, and foreign body sensation with increasing severity in recent years before admission. She had history of Steven-Johnson syndrome 17 years before. She had done skin and ear surgery to decrease the sequel of her SJS' symptoms. Heretofore, eyedrops and eye ointment moisturizer—artificial tears, cyclosporine and levofloxacin was used. In order to lessen the discomfort caused by her trichiasis, she also used bandage soft contact lenses.

Upon examination, her right eye's initial visual acuity (VA) was 20/25 could not be corrected. Furthermore, her left eye's initial VA was 20/80 could be corrected to 20/60 with correction lens S – 1.00 D. From non-contact tonometry examination founded that her intra ocular pressure on right eye (RE) was 17 mmHg, yet her left intra ocular pressure could not be measured. It was then founded keratinization on both of her eyelid margin with trichiasis and distichiasis. It

was also founded symblepharon and hyperemic conjunctiva, wide corneal scar, and neovascularization on both of her eyes as well (**figure 1**).

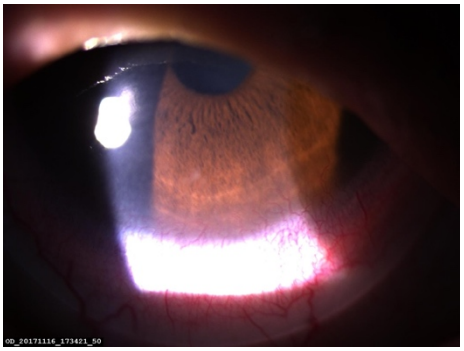
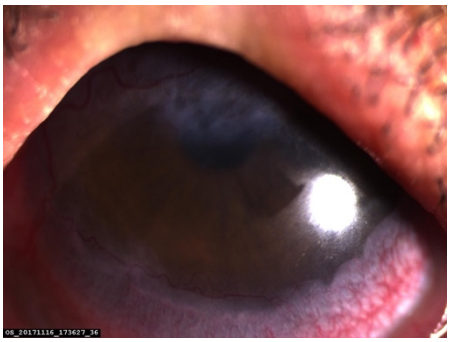
(a)  (b) 

Figure 1. Right (a) and Left (b) represented SJS' patient eye shows eyelid margin keratinization, distichiasis, symblepharon, conjunctival hyperemia and ciliary injection, corneal opacity and neovascularization

Hence, Jupiter® scleral contact lens was given to the patient following by lens fitting and the ideal fitting illustration, showed as good centrally coverage and movement along with ideal edge lift witgh no conjunctival indentation, was obtained (**figure 2**). The parameter of scleral contact lens which given to her was Base Curve 8.40 mm, Power S - 1.00 diopter, Diameter 14.7 mm for her RE, and Base Curve 7.70 mm, Power S – 1,75 diopter, dan diameter 14,7 mm for her left eye (LE). Then, her final VA was improved 1 Snellen level on the RE (20/25 to 20/20) and 5 line on the LE (20/80 to 20/25).

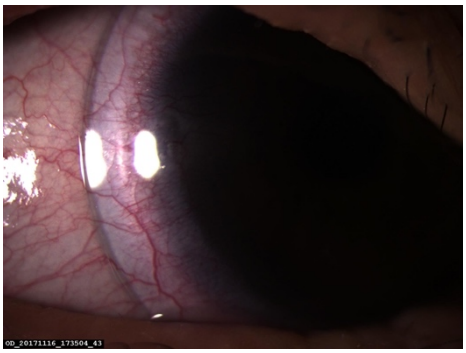
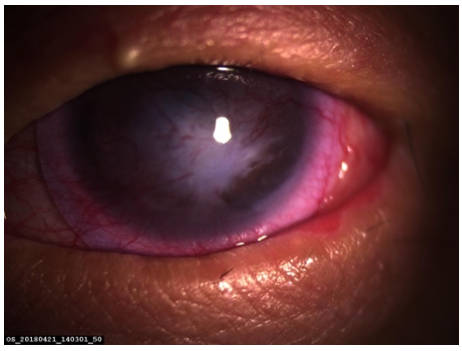
(a)  (b) 

Figure 2. Scleral contact lenses were fit ideally in Right (a) and Left (b) eye, with good centrally coverage and movement along with ideal edge lift with no conjunctival indentation

Following evaluation of the patient's complaint and physical examination was done after correction. It was founded that the lens position on central place with ideal lens movement. The patient's dry eye complaint was significantly decrease with more comfortable ocular sensation.

DISCUSSION

Steven-Johnson syndrome known as one of clinical spectrums of immunologic dermatobulous condition which has exceedingly high morbidity and mortality. Ocular surface recognized as one of main predilection sites of this disease, which could initiate blindless, both on acute and chronic phases.[1-3] SJS patients can also benefit from the use of scleral contact lenses. Ocular involvement on acute phases of SJS, caused by early keratinocyte apoptosis, secondary inflammatory effect, and epithelial loss on ocular surface. Previous studies revealed that ocular involvement founded in 50-88% SJS cases.[10-12] Ocular manifestation that are present at the beginning of SJS disease seems minor; however, the ocular manifestation may soon become worse.[13]

Acute ocular manifestation led to ocular chronic sequence in general, significant reduction of visual acuity followed later.[14] Chronic complication of SJS are symblepharon, ankyloblepharon, tarsal conjunctival scar which led to palpebral malposition, ectropion, entropion, trichiasis, distichiasis, atrophy of meibomian gland, lacrimal punctum occlusion, and keratinization on palpebral margin, tarsal and bulbar conjunctiva.[15] Hence, those can cause unpleasant pain and blindness, through corneal damage derives from chronic dysfunction of limbus stem cell.[16] Chronic corneal inflammation related to frequent blinking induce limbal stem cell dysfunction (LSCD), moreover neovascularization and scar tissue development on corneal as well.[16-19] Even after the acute disease episode has passed, neglected posterior palpebral margin keratinization during the chronic phase of SJS might damage the cornea and result in vision loss.[16]

Increased ocular surface lubrication and reduced inflammation were the objectives of adjunctive and supportive treatment for SJS patients. Comprehensive management consist of artificial tears, long-term ocular lubricant, punctal occlusion, cyclosporine, topical corticosteroid, and autologous eyedrops serum. Nevertheless, this supportive treatment still inadequate to increase quality of life patient with SJS. Topical ocular treatment generally beneficial for short-term management yet does not show adequate long-term improvement. Contrarily, several SJS patients experience subsequent corneal or ocular complications after treatment.[13,20]

It was known that scleral contact lens type rigid gas permeable (RGP) was an effective medical device for several ocular surface disruption treatment.[13,20] Previous cohort study by Rathi et al revealed that scleral contact lens type RGP also beneficial for ocular surface disruption treatment on SJS patient, especially for pain reliever and photophobia, furthermore it also could increase visual acuity as its secondary benefit. [21] All the research subject state that they have decreasing symptoms, although nearly the subjects develop debris under their lens. This debris led

them to repetitively remove, clean, and put on their scleral contact lens during period of application.[21]

Research from Tougeron-Brosseau et al also presents similar results, demonstrate that scleral lens uses for 6months could increase visual function, which rated from Visual Function Quality (NEI VFQ-25) questionnaire, and decrease ocular surface disease index (OSDI). These results are statistically significant (p=0.0001).[10] The mean score of NEI VFQ-25 pre and post contact lens usage are 25.1 ± 16.9 and 67.4±22.1. On the other hand, the mean score of OSDI also improve from 76.9±22.8 to 37.1±26.7. This study also revealed improvement of subjects' best corrected visual acuity (BCVA) to ≥ 2 Snellen level on 56.3% patient. Other studies also reported that 53-94.5% patient with ocular surface disease improve their visual acuity by using this lens.[10]

Stason et al, which also uses visual function questionnaire (NEI VFQ-25), stated that the most improvement are founded on ocular pain (97%) and dependency (81%).[22] The scleral contact lens application failure extremely low, it was founded on patient who are uncomfortable and uncooperative with scleral contact lens application.[22]

Case report from Campbell et al, revealed that improvement visual acuity, from 20/400 to 20/50, immediately post scleral contact lens application.[26] From this study, patient reported to be able to use this lens by themselves, in addition they were present significant improvement on ocular comfort and external findings. Scleral contact lens also gives satisfactory protection on ocular surface from palpebral scar tissue development and malposition of eyelashes without repeatedly epithelial disruption episode.[26] Fine et al also stated that scleral contact lens type RGP with high oxygen permeability generate significant improvement on patient. This was noticeable by their reduction in sign and symptoms, and artificial tears uses cessation.[27]

Apart from the main purpose of scleral contact lens application, stabilizes the corneal surface, it also useful for procedure of pre-reconstruction of ocular surface on visual blindness patient with SJS. Prior to visual acuity improvement, protection of ocular bulbi procedure with scleral contact lens could be firstly indicated to treat the persistent epithelial corneal defect. This procedure only can be done on eyes without widely symblepharon.[28]

CONCLUSION

In conclusion, scleral contact lens, especially type of RGP, should be considered as the SJS comprehensive treatment, specifically for patient who were already develop ocular complication. From this present case report founded how the patient had scleral contact lens fitted successfully for management of SJS ocular complication. They provided optimal visual quality, along with

prolonged use times and no adverse effects to the cornea. The lens might be improving the patient's function and quality of life.

Conflict of Interest

All Authors have nothing to disclose.

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