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Case Report

Management of Large Conjunctival Cysts in a Patient with Stevens-Johnson Syndrome: A Case Report and Review of the Literature

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Abstract
Stevens-Johnson syndrome (SJS) is a life-threatening mucocutaneous disease with various etiologies including drugs, infections, and malignancies. Ocular manifestations of SJS vary from the membrane, symblepharon formation, and epithelial defect in the acute phase to trichiasis, eyelid margin keratinization, and lacrimal duct obstruction in the chronic phase. A 13-year-old boy with a history of drug-induced SJS presented to our clinic complaining of a mass in the nasal side and inferior fornix of the right eye from 1 year ago. The mass-like lesion in the medial side of the right eye was accompanied by ankyloblepharon, symblepharon, and ptosis and limited ocular movement. Orbital imaging showed cystic lesions on the medial side of the right globe and the inferior fornix. Two large cysts were entirely surgically excised. Histopathologic investigation revealed conjunctival tissue with nonkeratinized epithelium and goblet cells. There was no sign of conjunctival cyst recurrence or symblepharon formation on the 6th-month follow-up. The inferior fornix achieved acceptable depth and the ocular movements became normal.

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Introduction

Stevens-Johnson syndrome (SJS) and toxic epidermal necrolysis (TEN) are at two ends of a spectrum of life-threatening mucocutaneous diseases [1]. They are specified by the severity of the body surface area (BSA) affected. SJS is defined as BSA involvement of less than 10%, while TEN is considered with more than 30% of BSA involvement. Of note, the overlap of SJS and TEN is between 10 and 30% of BSA. Regardless of classification, the SJS/TEN is characterized by keratinocyte apoptosis leading to denudation of the skin and mucosa including ocular surfaces [2].

The exact mechanism of SJS remains unknown; however, immunological T-cell mediated and non-immunological mechanisms have been proposed [1]. Various etiologies including drugs, infections, and malignancies are considered triggers. Common causative medications are sulfa derivatives, antiepileptics, and non-steroidal anti-inflammatory drugs. Infectious etiologies like mycoplasma pneumonia, herpes simplex, and malignancies including lung squamous cell carcinoma and Hodgkin's lymphoma are among the other triggers [3].

Ocular manifestations of SJS occur in almost half of the patients with SJS and range from conjunctival hyperemia, membrane, and subsequent symblepharon formation and epithelial defect in the acute phase to cicatrizing conjunctivitis, trichiasis, eyelid margin keratinization, and lacrimal duct obstruction in the chronic phase [4]. Orbital or conjunctival cysts are reported occasionally as a late sequela of SJS. Cysts may originate from the conjunctiva or the lacrimal glands. Smaller cysts may be asymptomatic but larger ones are symptomatic and may restrict ocular motility and cause SJS signs to exacerbate [5, 6]. Herein, we report a patient with multiple conjunctival cysts as a late sequela of SJS. We also performed a literature review on the management of orbital and conjunctival cysts in patients with SJS/TEN. The CARE Checklist has been completed by the authors for this case report, attached as online supplementary material (for all online suppl. material, see https://doi.org/10.1159/000533648).

Case Report

A 13-year-old boy presented complaining of a mass in the nasal side of the right eye from 1 year ago. He had no history of ocular surgery and trauma. He had a history of indomethacin- or cefalexin-induced SJS for the last 3 years. There was no other past medical or surgical history. The best-corrected visual acuity was counting fingers from 2 m in the right eye and counting fingers from 6 m in the left eye.

Ocular examination revealed a mass-like lesion in the medial side of the right eye accompanied by ankyloblepharon, symblepharon, and ptosis (Fig. 1a). Symblepharon and poliosis were observed in both eyes. Ocular movements were limited secondary to symblepharons. Severe keratinization of eyelid margins was also evident in both eyes. Pupils were equal and reactive with no relative afferent pupillary defect. On slit lamp examination, cicatrized changes of eyelids were observed. Also, superior and inferior symblepharon, dry eye, two conjunctival cysts, and diffuse punctate erosion on the corneal surface were observed in both eyes. Moderate corneal opacity and vascularization were evident in both eyes. Other ocular examinations were within normal limits. Computed tomography scan of the orbit revealed a large cystic lesion on the medial side and a smaller cyst inferonasal to the right globe (Fig. 1b, c).

The patient was scheduled to excise the cysts. Through an inferomedial conjunctival incision, the medial conjunctival cyst was meticulously dissected from the medial rectus as well as the upper/lower eyelids, and the cyst was entirely removed. In addition, the inferior

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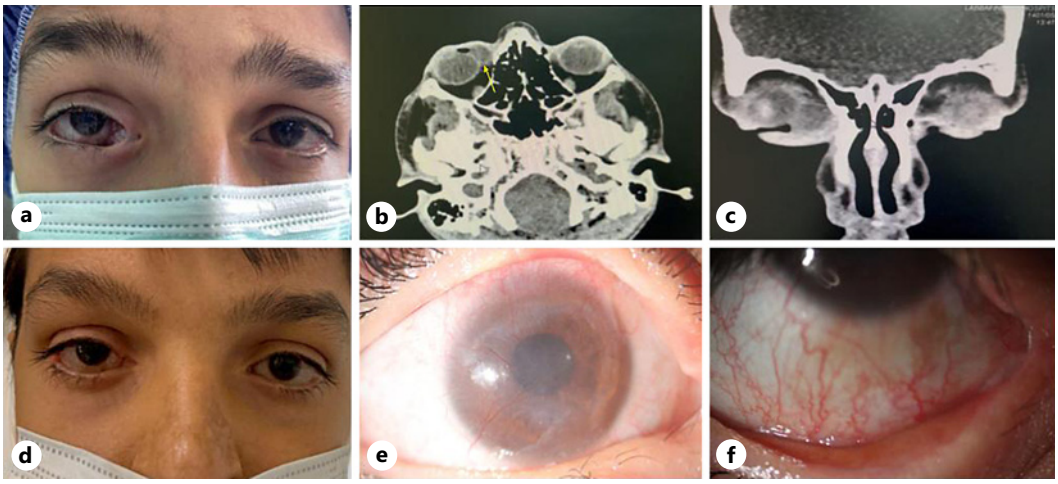

Fig. 1. a Preoperative image reveals two conjunctival masses located behind the medial canthus accompanied by ankyloblepharon and inferior fornix. **b** The axial plane of the computerized tomography (CT) scan shows an isodense lesion located extraconally in the medial portion of the right eye. **c** The coronal plane of the CT scan shows an isodense well-defined lesion in the inferomedial of the right eye. Postoperative images on month 4 show no sign of conjunctival mass or ankyloblepharon (**d**), corneal vascularization (**e**), and an acceptable fornix depth (**f**).

cyst was excised through a directly forniceal conjunctival incision. A 5 * 5 cryopreserved amniotic membrane was placed in the place of resected cysts, and a fornix conformer was put in place (Fig. 2). The patient used topical antibiotic for a week and steroid for 3 weeks, as well as lubricant therapy. The fornix conformer was removed 2 weeks later, and the miniscleral design (MSD) lenses were fitted for the patient. Miniscleral lens-corrected visual acuity reached 20/50 in both.

Histopathologically, the mass showed conjunctival tissue with a cystic structure in substantia propria, lined by a double layer of nonkeratinized epithelium with goblet cells. The patient was followed up 6 months after surgery and there was no sign of orbital cyst recurrence or symblepharon formation. The inferior fornix had acceptable depth and normal ocular movements (Fig. 1d–f). Miniscleral-corrected visual acuity reached 20/40 using the Snellen chart. Informed consent including publication of photographs in medical journals was obtained from the parents of patient.

Discussion

We presented an SJS patient with two conjunctival cysts extending into the orbit without previous ocular surgery or trauma which was successfully treated. Orbital cysts can be classified by their manifestation, pathogenesis, or histopathology. They are primary or secondary to trauma, surgery, or inflammation [7]. Conjunctival epithelial cysts are simple cysts lined by conjunctival epithelium containing goblet cells. Primary conjunctival cysts are usually located in the supranasal side of the orbit without proptosis or globe displacement. However, larger cysts can limit eye movement, displace the globe, and induce refractive error [7, 8]. In contrast, the location of secondary conjunctival cysts following trauma or surgery depends on the type of surgery or site of trauma.

Conjunctival cysts are relatively rare ocular manifestations in the chronic phase of SJS/TEN. However, the conjunctival cysts may seriously affect patients with SJS. Besides the

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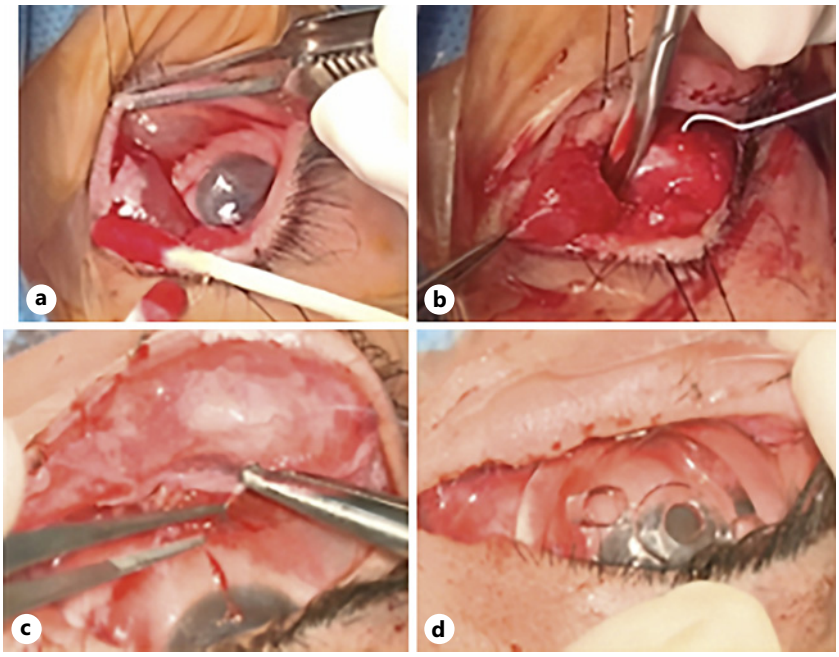

Fig. 2. Images demonstrating the surgical steps of cyst excision. **a** The medial and inferior cysts are visible. First, the medial cyst is meticulously dissected from the medial rectus as well as the upper/lower eyelids (**b**). A 5 * 5 cryopreserved amniotic membrane was placed in the place of resected cysts (**c**) and a fornix conformer was put (**d**).

cosmetic issues, the larger cysts may exacerbate the dry eye, cause Dellen formation, or prevent implantation of miniscleral lenses like MSD. MSD lenses are among the best treatment options for patients with SJS due to significant visual improvement and reducing the microtrauma caused by eyelid margin irregularity and keratinization [5].

To the best of our knowledge, to date, five studies reported conjunctival or orbital cysts in patients with SJS (Table 1) [5, 6, 9–11]. The epithelial cysts may originate from the conjunctiva or the lacrimal glands. It is assumed that chronic inflammation in SJS may cause adhesions between the palpebral and bulbar conjunctiva with subsequent entrapment of epithelial cells and cyst formation. Moreover, cyst enlargement may be the result of mucus secretion by conjunctival epithelium. The irregularities and cysts prevent tears from reaching the conjunctival surface [6].

Surgical management of conjunctival cysts consists of two approaches, including complete intact excision versus marsupialization, which means unroofing the cyst. While complete cyst resection using meticulous resection remains the standard technique, preserving the intact walls is not always possible. Memarzadeh et al. [9] advocated that marsupialization in large conjunctival cysts located in the inferior fornix could preserve the epithelial lining to be used in fornix reconstruction. However, marsupialization may not be a good choice in deeper orbital/conjunctival cysts. In our patient, despite confirmed adhesion of the cyst to the eyelids and medial rectus sheet, complete cyst resection was possible for both adjacent cysts, and the inferior fornix was reconstructed with AMT. Because of the inflammatory process, in patients with SJS, the conjunctival cysts may have a more confirmed attachment to the adjacent tissue and extraocular muscles. So meticulous dissection is needed to complete the excision of such conjunctival/orbital cysts. However, in superficial epithelial conjunctival cysts with inadvertent cyst rupture or a high probability of rupture, marsupialization can be alternatively used.

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Table 1. Summary of studies reporting conjunctival and orbital cysts in patients with SJS

Study	Age	Sex	Laterality	Manifestation	Time of SJS	Cause of SJS	Treatment	F/U duration	Outcome
Singh et al. [5] (2008)	11	M	OU	Conjunctival cyst at the medial of inferior fornix	3 years	–	Observation	–	–
Memarzadeh et al. [9] (2006)	17	M	OU	Inferior anterior orbit and lower eyelids cysts	1 year	Tetracycline	Marsupialization	1 year	No recurrence
Desai et al. [6] (1992)	10	F	OD	Superior anterior orbit. conjunctival origin	5 years	Phenobarbital Erythromycin Acetaminophen Aspirin	Cyst excision	–	No recurrence
Goodglick et al. [10] (1992)*		F	OU	Palpebral conjunctival cysts	NA	Allopurinol	–	–	–
Harris et al. [11] (1983)*				Lacrimal gland cyst		Not reported	Marsupialization of the wall	–	–
The present study	13	M	OD	Conjunctival cyst, adjacent to the medial rectus muscle	3 years	Indomethacin, cephalexin	Cyst excision	6 months	No recurrence

SJS, Stevens-Johnson Syndrome; OU, oculus uterque; OD, oculus dexter; OS, oculus sinistra.
*The full-text was not available.

In conclusion, we presented the surgical management of an SJS patient with two large conjunctival cysts located in the nasal and inferonasal orbital cavities. Due to lid deformity, ptosis, and movement limitation, the patient underwent surgical cyst resection and inferior fornix reconstruction. Cyst excision benefited our patient in terms of miniscleral lens fitting, resolution of lagophthalmos, and dry eye disease improvement.

Statement of Ethics

Written informed consent was obtained from the parents of the patients for publication of the details of their medical case and any accompanying images. Ethical approval is not required for this study in accordance with local or national guidelines.

Conflict of Interest Statement

The authors declare no conflict of interest.

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Author Contributions

Concepts: Mohammad Ali Javadi, Mohammad-Mehdi Sadoughi. Design and definition of intellectual content: Kiana Hassanpour, Amirreza Veisi. Literature search and manuscript preparation: Kiana Hassanpour, Sadid Hooshmandi, Vahid Movafaghi, Farideh Langari. Clinical studies and experimental studies: Kiana Hassanpour, Sadid Hooshmandi. Data acquisition: Kiana Hassanpour. Manuscript editing and manuscript review: Mohammad Ali Javadi, Mohammad-Mehdi Sadoughi, Amirreza Veisi.

Data Availability Statement

All data generated and analyzed during this study are included in this article. Further inquiries can be directed to the corresponding author.

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Case Rep Ophthalmol 2023;14:528–534
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