Bilateral conjunctival retention cysts in the aftermath of Stevens-Johnson syndrome

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In this case report, we describe the rare occurrence of bilateral conjunctival retention cysts in a child with Stevens-Johnson syndrome. The case was managed conservatively as there were no functional or cosmetic problems.

Key words: Bilateral, conjunctival retention cyst, Stevens-Johnson syndrome

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Extensive ocular surface scarring is the common denominator in Stevens-Johnson syndrome (SJS) causing complications such as symblepheron, dry eye, ocular surface keratinization, lacrimal duct obstruction and lid deformities.[1] Conjunctival and orbital cysts were reported occasionally.[2] We describe the occurrence of bilateral conjunctival retention cysts in a child with SJS.

Case History

An 11-year-old male child was referred for epiphora and bilateral conjunctival swelling. He was admitted in a hospital and treated for SJS after an attack of brief fever, three years

before. The hospital notes recorded supportive management with intravenous fluids, antibiotics and an advice to avoid analgesics, anti-inflammatory agents, penicillin and sulpha. Though ocular involvement was noted there were no details about its management. On examination, he had uncorrected visual acuity of 20/20 in both eyes. Anterior segment examination revealed bilateral palpebral conjunctival scarring, patchy obliteration of inferior fornices and cicatricial occlusion of all puncta. An ovoid, translucent conjunctival cyst was seen at the medial end of each inferior fornix, behind the cannalicular area. They were easily prolapsed by everting the eyelid and applying upward pressure [Figs. 1 A and B]. The cysts seem to extend from adjacent palpebral conjunctival scarring. A cilium was incorporated in the left cyst. The lid margins were otherwise normal without significant keratinization. The corneas were clear with no significant staining. The tear meniscus was adequate in both eyes. Tear film breakup time and the Schirmer's test values were within normal limits. There was no swelling over the lacrimal sac areas and no regurgitation was seen on pressure over the sacs. The ocular motility was normal. Since the ocular surfaces were healthy despite the conjunctival scarring and the cysts did not cause cosmetic or functional problems, we decided to observe the patient and the same was explained to the parents.

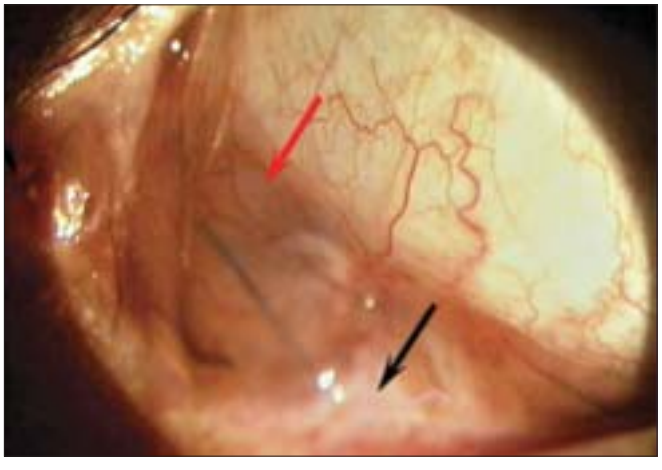

Figure 1A: Conjunctival linear scarring, adjacent translucent cyst and cilium within

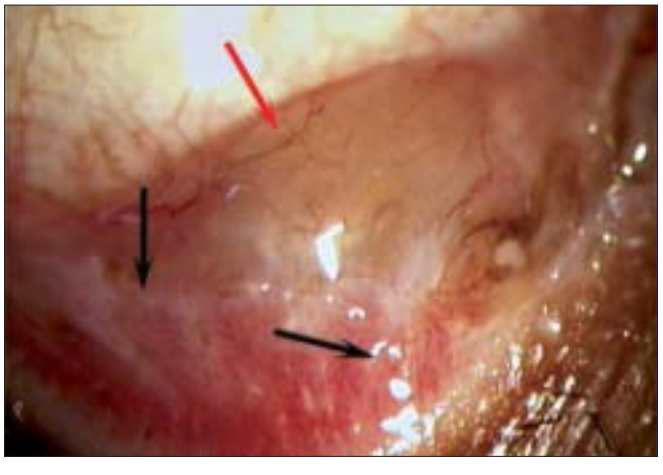

Figure 1B: Occluded punctum and the cystic mass

Discussion

Conjunctival cysts may be congenital or acquired. Acquired conjunctival cysts can occur due to sequestration of the conjunctival cells adjacent to scars after penetrating trauma, surgeries involving conjunctival and tenon's fascia manipulations like strabismus surgery or even after sub-tenon's injection of anesthetic agents.[3,4] Cicatricial ocular inflammations are another common source for conjunctival cysts. The extensive surface inflammation with adhesions between the tarsal and bulbar conjunctival surfaces can cause sequestration of epithelium beneath the surface with the formation of retention cysts, in ocular surface inflammatory conditions.[5] In SJS extensive surface denudation and inflammation in the acute phase can lead to such adhesions between the healing conjunctival surfaces. Even in the late cicatricial phase, ongoing epithelial hyperproliferation with inflammatory cell infiltration in the substantia propria has been demonstrated in the conjunctiva.[6] This can lead to cyst formation from the proliferating sequestrated epithelial cells as in our case. This inclusion can be further aided by practices like breaking the conjunctival adhesions with glass rods during the acute phase. The presence of cilium in one of the cysts could have occurred by this mechanism. Chronic ocular surface inflammation is also attributed to the formation of cysts in conditions like pterygium, vernal keratoconjunctivitis.[7] In SJS ongoing ocular surface inflammation has been well established.[8]

While smaller cysts can be of cosmetic problems, larger ones can act like space-occupying lesions and restrict ocular motility. Surgical interventions like excision and marsupialization were suggested for larger cysts.[5] We have chosen a conservative approach, as they were cosmetically insignificant, with good lid closure and a stable ocular surface. Lacrimal drainage system obstruction causing bilateral dacryocystoceles in SJS have been reported and managed with dacryocystorhinostomy with silicone tube insertion.[9] We opted to manage the epiphora also conservatively to maximize the protective effect of punctal occlusion in maintaining a healthy ocular surface as reported by Kaido *et al*.[10]

Stevens-Johnson syndrome can result in devastating ocular surface scarring and keratinization in its severe form. This report extends the spectrum of minor long-term ocular complications associated with SJS. Caution during procedures like glass rodding in acute phase and controlling inflammation in the late phase can reduce this complication.

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