Novel surgical technique for the management of partial cryptophthalmos

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We report a case of a 2-month-old baby with bilateral nonsyndromic partial cryptophthalmos presenting with upper eyelid incomplete development and fusion to the cornea with resultant inability to close the eyes. He was managed successfully with bilateral upper lid reconstruction with composite graft using maternal skin and oral mucous membrane, amniotic membrane, and donor scleral graft. After this one stage surgery, lids were well-formed, and the patient was able to close both eyes, thus achieving good anatomical, functional, and cosmetic outcome.

Key words: Congenital symblepharon, lid reconstruction, partial cryptophthalmos

Cryptophthalmos (Greek, Kryptos [hidden] + ophthalmos [eye]) was first described by Zehender and Manz in 1872.[1] It is characterized by partial or complete coverage of eyeball with skin and may occur in isolation or as a part of Fraser syndrome.[1] Cryptophthalmos presents as a challenge even in the present times. Facial dysmorphism, near absence of vision and inability to close eyelids causing severe ocular surface insult are the major issues which need to be addressed. Often, they have microphthalmos and systemic abnormalities such as syndactyly and urogenital malformations.[2] Cutaneous symblepharon, with absence of tissue, makes the lid reconstruction difficult and mandates use of multiple grafts. The goal of surgery is to release the symblepharon, repair the coloboma and reconstruct eyelids, visual prognosis, however remains bleak. Most of the procedures described in the literature are multi-staged and involve considerable donor site morbidity.[3] We describe a quadruple graft, single-stage technique using maternal donor tissue and allografts, performed bilaterally, hence restoring complete anatomical and functional correction after single surgery with no donor site morbidity for the patient.

Case Report

A 2-month-old Indian male infant presented with bilateral congenital cryptophthalmos involving entire upper lids. The upper lid eyelashes and eyebrows were absent, and a tongue of temporal hair extended onto the forehead. The forehead skin swept down to form a cutaneous symblepharon with the upper one-third of the cornea. The child was unable to close the eyes with resultant desiccation of the ocular surface. Lower lid skin was deficient with mild ectropion [Fig. 1]. Cornea was opaque, and ocular motility restricted due to symblepharon. On ultrasound, posterior segment was normal. The child had perception of light and responded to the flashlight of the camera. The condition was bilaterally symmetrical. There was no associated facial or systemic abnormality. The child was diagnosed to have congenital bilateral cryptophthalmos – congenital symblepharon variant.[1]

The patient underwent bilateral lid reconstruction under general anesthesia. For reconstructing the anterior lamella, skin graft was chosen, for the tarsal plate, a donor scleral graft was chosen, for the conjunctiva replacement, mucous membrane was used, and amniotic membrane was used to cover the grafts on the conjunctival aspect [Fig. 2]. A 26 mm × 30 mm left retro-auricular skin graft and 20 mm × 26 mm oral mucosal graft was harvested from the mother. Each of these was divided into half for use in both eyes. After detaching the cutaneous symblepharon from the ocular surface, the upper lid skin was incised about 3 mm from the edge and the tissues explored. Some remnants of the levator palpebrae superioris (LPS) muscle were isolated. A 26 mm × 15 mm skin graft was sutured along the entire length of the gap created due by the skin incision [Fig. 3a]. The LPS remnants were sutured to donor skin as well. An 18 mm × 6 mm scleral graft was used to reconstruct the tarsus. There was minimal remnant of conjunctiva in the rim at the lid margin side to which the scleral graft was anchored. A 20 mm × 13 mm maternal mucous membrane graft was sutured to the superior part of the scleral graft and superiorly in the forniceal area thus creating an upper fornix. The entire ocular surface and newly constructed upper lid was lined by fresh amniotic membrane and sutured in place [Fig. 3b]. An 18 mm size symblepharon ring was placed and suture tarsorrhaphy performed. Postoperatively, the patient was on oral antibiotics and steroids for 1-week, followed by a tapering dose of steroids for another week. The graft was taking up well at the end of the 1st week [Fig. 3c]. Topical medication included antibiotics, steroids and lubricating eye drops, as well as antibiotic eye ointment. Tarsorrhaphy was released after 1-month and the symblepharon ring removed. All grafts healed well and reconstructed lid covered the entire eye at the last follow-up 18 months after surgery. Lids were mobile, thus providing complete functional and anatomical restoration [Fig. 4a and b].

Discussion

Cryptophthalmos is a challenging scenario with no set guidelines for its management. The goal is to restore cosmesis and prevent further ocular surface insult. The choice of surgery depends on the severity of the condition, age of the patient and surgeon's preference. Brazier *et al*. described a Mustarde's lower lid rotation flap performed on a 6 day old with satisfactory results.[1] This was a 2 staged procedure requiring a flap division after 3 weeks. This technique provided upper lid eyelashes. However, lid sharing from lower lid was not preferred in the

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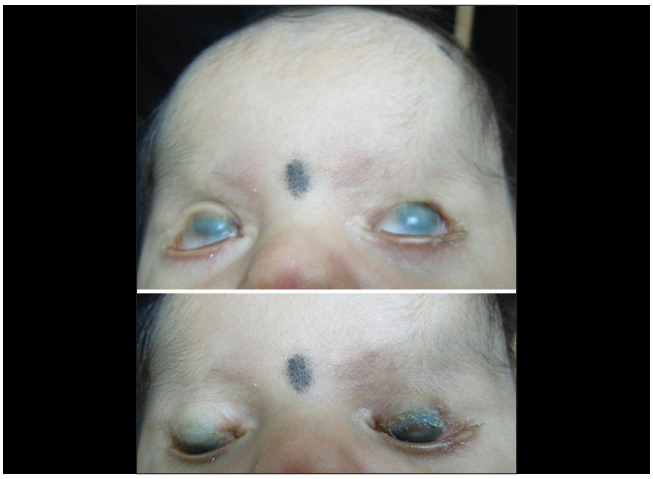

Figure 1: Bilateral upper lid cryptophthalmos with lower lid ectropion and cutaneous symblepharon involving superior part of the cornea with inability to close eyelids with ocular surface compromise

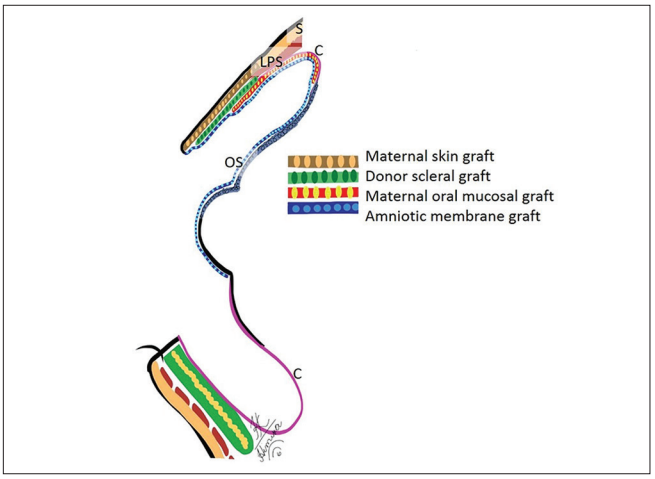

Figure 2: Schematic representation of grafts. S-upper lid/brow skin, levator palpebrae superioris remnants, C-conjunctiva, OS-raw ocular surface

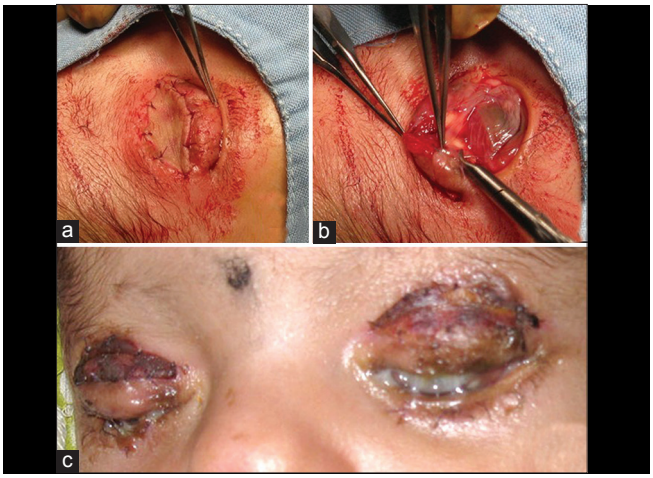

Figure 3: (a) Maternal skin graft. (b) Amniotic membrane graft lining the ocular surface. (c) Bilaterally reconstructed lids, after surgery

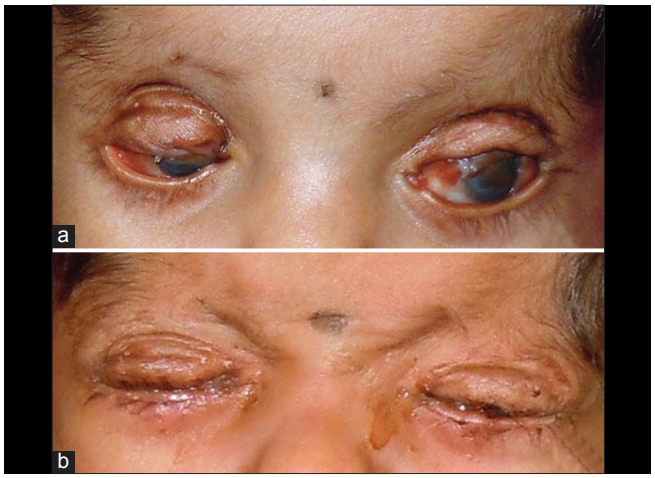

Figure 4: One-year after surgery (a) reconstructed upper lids (b) proper lid closure with no lagophthalmos

current case due to preexisting lower lid skin shortening and ectropion. Subramanian *et al*. have advocated Mustarde's lid switch with amniotic membrane lining.[3] They also recommend lower lid tarsoconjunctival advancement with skin graft for anterior lamella. Weng described a sandwich conchal graft with oral mucosa in a 3-staged procedure to achieve good functional and cosmetic outcome.[4] All these procedures are multi-staged and involve significant donor site morbidity.[5]

Our technique achieved good anatomical, functional, and cosmetic rehabilitation in a single surgical sitting, thus avoiding the need of subsequent surgeries. Maternal tissue provided a good match, and avoided donor site morbidity in the patient. Mother's skin was chosen as there was reluctance from the family members to taking a graft from the child's body. Although we did not perform human leukocyte antigen (HLA) matching, we expected close HLA matching of the mother's graft. Maternal skin has been successfully used in patients with generalized skin disorders with no scope of autograft harvesting.[6] Mucous membrane was chosen as it provides a moist surface akin to the conjunctiva. The prognosis of multiple layered free grafts is generally considered guarded. However, the placement of the grafts in this surgery was such that every layer was in direct contact with some amount of recipient tissue which provided the blood supply and nourishment for the graft to survive. The LPS remnants rendered motility to the lids and enabled proper blinking and good functional results.

Single-staged bilateral surgery using maternal graft is a viable option for lid reconstruction in partial cryptophthalmos, especially in infants. It obviates the need of further procedures and achieves quick and complete ocular surface protection. Maternal graft is especially useful when it is not feasible to harvest enough autologous donor tissue.

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