



Surgical management of large bilateral epibulbar dermoids with autologous oral mucous membrane transplantation

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ABSTRACT

Purpose: To report the surgical management of extensive epibulbar dermoids with autologous oral mucous membrane transplantation.

Observations: While rare, extensive dermoids that encroach upon the visual axis carry a poor prognosis. We report the case of a 7-week old premature male infant who presented with large bilateral epibulbar dermoids obscuring the visual axis. He was treated first with sequential bilateral optical iridectomies under the clearest corneal areas, followed several months later by sequential dermoid excision and amniotic membrane transplantation in each eye. He subsequently underwent autologous “simple” oral mucosal epithelial transplantation (SOMET) as well as strabismus surgery. **Conclusions and Importance:** Here we present the first case, to the best of our knowledge, of the use of SOMET in managing post-operative pseudopterygium following dermoid excision. To our knowledge it is the also the first application of this technique in a young pediatric patient. A good clinical outcome may be achieved with SOMET, which may offer a minimally invasive alternative to other traditional modalities.

1. Case presentation

A 7-week-old male infant presented for evaluation of bilateral epibulbar dermoids (Fig. 1A and B). He was born prematurely at 31.5 weeks as a fraternal twin, complicated by intrauterine growth restriction. Past medical history was significant for chromosome 1p32.2 deletion on microarray, but there was no family history of eye disease. On exam, he had reaction to light in each eye, and large bilateral Grade II epibulbar dermoids, classified based on involvement of nearly the entire cornea obstructing the visual axis with invasion down to deep stroma but without penetration into the anterior chamber. His CT scan confirmed a poorly developed nasolacrimal system bilaterally; there was no visualized nasolacrimal duct on the left side. Physical exam showed broad nasal bridge, bifid nose, and midface hypoplasia. A multidisciplinary approach was undertaken, involving cornea, glaucoma, oculoplastics, and pediatric ophthalmology. The option of dermoid excision with corneal transplantation was reviewed, but based on his health issues, the decision was made to start with optical iridectomy under the clearest corneal areas to permit visual development.

At 3 months chronological age, approximately 51-weeks post-conceptual age, he was taken for examination under anesthesia (EUA)

(Fig. 1C and D). Due to his mucous pooling and poorly developed lacrimal system: there was concern for increased risk of post-operative infection, so optical iridectomy was performed only in the left eye. Post-operatively, he did well and so underwent optical iridectomy in the fellow eye one month later. At 5 months of age, he continued to have poor visual behavior with poor fixation and poor tracking of objects (right eye more than the left).

Dermoid removal with option of cornea transplant and possible limbal stem cell transplantation (LSCT) was again reviewed with the parents. While the patient was not a candidate for conjunctival limbal autograft (CLAU) or autologous simple limbal epithelial transplantation (SLET), due to the large dermoid (and limited limbal stem cell reserve) in the fellow eye, the options of keratolimbal allograft (KLAL) alone, or combined KLAL with a living-related conjunctival limbal allograft (Ir-CLAL; “Cincinnati procedure”) were discussed. The parents were, however, concerned about the higher risks associated with transplantation, and especially systemic immunosuppression. The resources for cultivated oral mucosal epithelial transplantation [COMET] and cultivated limbal stem cell transplantation [CLET] are not available at our institution.

At age 8 months, the patient underwent extensive resection of the

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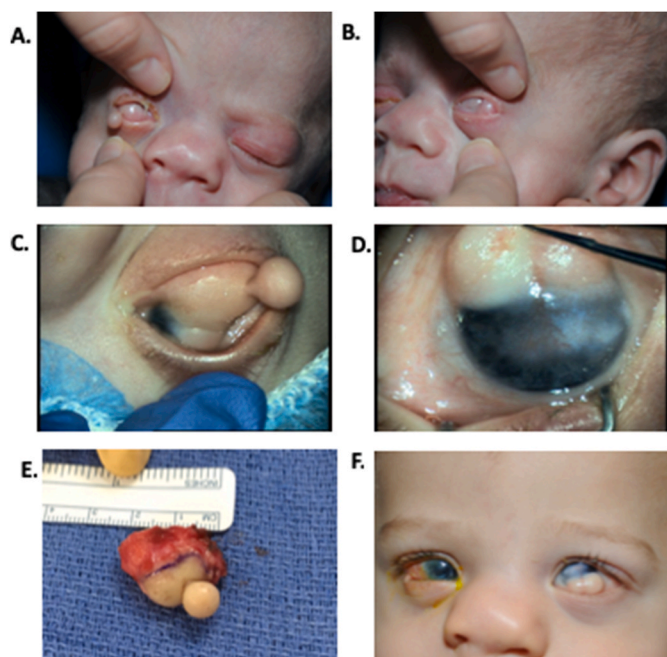


Fig. 1. External photos of bilateral corneal dermoids at the time of first examination in the NICU at age 31.5 weeks gestation, right eye (A), left eye (B). Intraoperative photos from examination under anesthesia, right eye (C) and left eye (D). E. Intraoperative specimen. F. External photo at post op month 1 from bilateral limbal dermoid excision.

right orbital and epibulbar dermoid, and ocular surface reconstruction with amniotic membrane transplantation (Fig. 1E). The visual axis was surprisingly clear post-operatively in the right eye, with good fix and follow (Fig. 1F). However, he soon developed a right eye preference with worsening visual behavior in the left eye (without fix or follow, and esotropia), and this led to the decision to perform the same procedure on the left eye. Subsequently, both corneas were well epithelialized with clear visual axis; the patient had less objection to occlusion and improved esotropia.

At his four month post-operative visit, the right eye had reaction to light with poor fix and follow, and the left eye had fix and follow with nystagmus. Examination showed increased conjunctival inflammation with encroachment of pseudopterygium onto the visual axis bilaterally. His pediatric ophthalmologists noted progressively worsening esotropia, nearly 70 prism diopters, with eccentric fixation in the left eye and poor fixation in the right eye. Given pseudopterygium recurrence, the option of Mitomycin C (MMC) and LSCT was again considered, but ultimately the decision was made to proceed with (1) repeat bilateral ocular surface reconstruction with amniotic membrane transplantation, (2) autologous “simple” oral mucosal epithelial transplantation (SOMET), and (3) simultaneous strabismus surgery to correct the esotropia.

In brief, a combined approach with oculoplastics was undertaken to first dissect the orbital mass, followed by dissection of the pseudopterygium and scar from the surface of the cornea. Once it was felt that the ocular surface dissection was sufficient, attention was turned to the oral mucous membrane harvesting. Topical anesthetic was administered to the labial mucosa and a Hessberg Barron 9.0mm trephine was used to trephine the central portion to 250 μ m. A Crescent blade was then used to dissect outwards and harvest the labial mucous membrane along with a 15 blade and Westcott scissors. The labial mucosa was soaked in 5% betadine for 1 minute to decrease the risk of infection. Amniotic membrane was draped over the cornea and then the mucous membrane was cut into a $\frac{3}{4}$ portion, which was placed around the limbus of the cornea and sutured in place with 9-0 vicryl.

At his post-operative month 1 follow up visit after SOMET, age 16 months, he had clear visual axis and improving visual behavior with

reaction to light in the right eye and good fix and follow in the left eye (Fig. 2A). The improved ocular surface allowed for better visualization of the posterior segment, revealing anomalous left optic nerve suspicious for optic nerve coloboma. The visual axis remained clear at POM3 (Fig. 2B) and at his most recent follow up visit at age 27 months, 1 year after SOMET, fixation was UCsUM in the right eye, UCSM in the left eye. The visual axis remains clear (Fig. 2C and D). He has persistent esotropia and amblyopia of the right eye but with good visual function in the left eye. His refraction is -3.00 right eye, -4.75 left eye.

2. Discussion

Limbal dermoids are the most common choristomas, often located inferotemporally. Corneal dermoids that encroach on the visual axis carry a poor prognosis; induced astigmatism and growth of these lesions can lead to anisometropic or deprivational amblyopia. Limbal dermoids have been traditionally classified into three grades: grade I as the most superficial and smallest (less than 5mm in size), grade II with lesions that extend into the corneal stroma and Descemet’s membrane, and grade III lesions which cover the entire cornea and can penetrate into the anterior chamber. The preferred treatment depends on the size and location of the lesion.¹ A more recent validated grading system has proposed revising the traditional limbal dermoid scoring, to account for the area of cornea and conjunctival involvement, as well as for the surface shape of the dermoid.²

Smaller, superficial lesions can be treated with simple excision.^{3,4} Excision of larger dermoids require more extensive reconstruction and possible lamellar or penetrating keratoplasty, which carry their own challenges in pediatric patients.⁵⁻⁹ Post-operatively, large dermoid excision has been associated with scar formation, pseudopterygium, and corneal neovascularization. Adjunctive procedures at the time of surgical excision have been developed to prevent these complications. The use of topical MMC has been shown to be protective against pseudopterygium formation, although long-term data is not available.¹⁰

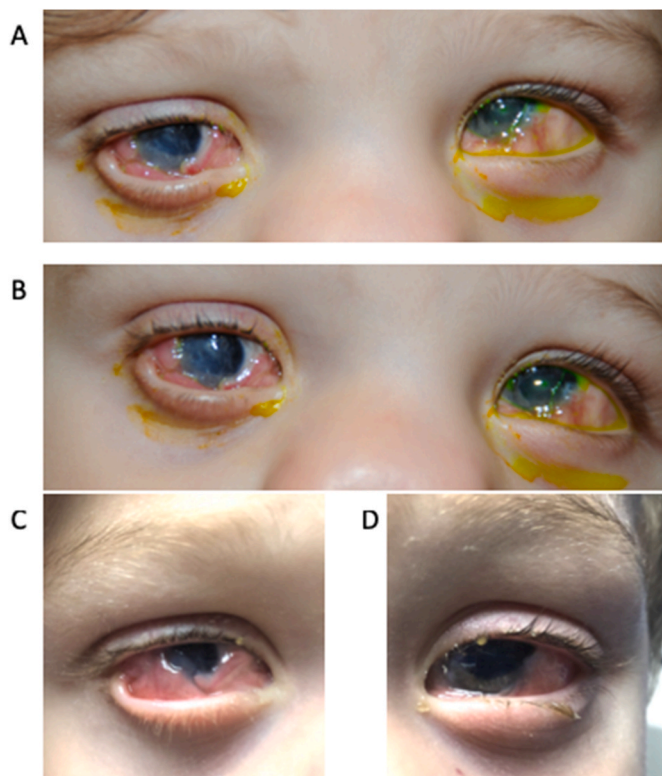


Fig. 2. External photos at one month (A), 3 months (B), and one year (C, Right eye, D, left eye) post-operatively from SOMET.

Topical MMC also has additional risks of scleral thinning, infectious scleritis, and globe perforation, which must be considered, particularly in a young pediatric patient. Amniotic membrane has anti-inflammatory and anti-angiogenic properties: more recently it has been described in ocular surface reconstruction after epibulbar dermoid excision with good results.^{11,12} In cases of severe limbal stem cell deficiency (LSCD), LSCT can be considered. Autologous LSCT has been described in patients undergoing dermoid excision with good results.¹³ However, this is not possible in cases of large bilateral corneal dermoids, as in our patient. Allograft LSCT is the gold standard in bilateral LSCD, but necessitates systemic immunosuppression. More recently, SOMET, direct transplantation of autologous (circumferentially-trephined) oral mucosal graft for ocular surface reconstruction, has been reported as an effective treatment for severe LSCD in patients with Steven Johnson Syndrome (SJS).¹⁴ In this case series of four SJS patients, all patients showed improved vision at 4 months post-operatively with successful reconstruction of the ocular surface. SOMET has the advantage of precluding the need for systemic immunosuppression (as in cases of KLAL or Ir-CLAL), and can be used in cases of bilateral corneal disease.

3. Conclusions

To our knowledge, our patient is the first application of autologous SOMET in the case of limbal dermoid, and the first application of autologous SOMET in a young pediatric patient. When SOMET is used in young children, one must consider that oral mucosa harvesting can risk feeding difficulties and children should be monitored closely for proper nutrition and healing. Further follow up data is needed regarding long term outcomes and the survival of SOMET grafts, but in cases of LSCD, our case provides evidence that SOMET may offer a minimally-invasive alternative to allograft limbal stem cell transplantation, especially where systemic immunosuppression is not feasible (or undesired by the patient or family).

Patient consent

Consent to publish this case report has been obtained from the patient(s) in writing.

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Conflict of Interest statement

Dr. Louise Mawn is a consultant for Horizon Therapeutics, but they did not provide any financial contribution to this work, nor is she a consultant to them for an area of ophthalmology related to this case report

The authors certify that they have no conflict of interests to disclose.

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