CASE REPORT

Childhood bilateral limbal stem cell deficiency: long-term management and outcome

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SUMMARY

A 6-year-old boy presented in 1998 with a 1 month history of lime injury in both eyes. Grade 4 chemical injury (Roper-Hall classification) progressed to total limbal stem cell deficiency in both eyes. Between July 2000 and July 2009, he cumulatively underwent 11 limbal stem cell transplantation surgeries, either direct or cultivated from either his father or mother, and 3 corneal transplants in both eyes. Each of the procedures led to a transient clearing of his visual axis to give vision in the range of 20/60 to 20/100 for a period of 1–3 months after which it failed. He finally underwent Boston keratoprosthesis implantation and now enjoys 20/20 vision with stable optic disc and visual field at a follow-up of 41 months. Timely intervention in childhood helped in prevention of amblyopia in this patient. Perseverance by the patient, his relatives and treating ophthalmologist is very important.

BACKGROUND

By virtue of a follow-up of 12 years, this case highlights the fact that management of childhood chemical injuries poses both a strategic and therapeutic challenge. Early and repeated interventions are sometimes necessary in children to maintain or extend the period of visual axis clarity to prevent the onset of amblyopia, before more complex and definite treatments like limbal allografting and Boston keratoprosthesis are in place.

CASE PRESENTATION

A 6-year-old boy presented to us in September 1998 with an injury with lime powder in both eyes 1 month ago. He underwent local ophthalmic consultation where a thorough eye wash with normal saline was performed in both his eyes and he was prescribed some eye drops, the details of which were unavailable with him at presentation.

At his first visit with us, he had a best-corrected visual acuity (BCVA) of counting fingers (CF) at 1 m and 20/100 in the right eye (OD) and left eye (OS) respectively. The clinical picture in both eyes was similar. Conjunctiva was congested, cornea showed total epithelial defect with stromal oedema severe enough to obscure the iris details, anterior chamber was deep and the lens was clear. Fundus view was hazy but a good fundal glow could be appreciated. B-Scan ultrasonography of both eyes was normal. Based on the above findings, a clinical diagnosis of Grade 4 chemical injury (Roper-Hall classification)¹ in both eyes was made. He was put on topical antibiotics, lubricants and betamethasone 0.1% eye drops six times a day.

On treatment, the ocular surface progressively healed with 360° pannus formation over the next 3 months. He presented to us in December 1998 and then in July 2000 with total limbal stem cell deficiency (LSCD) in both eyes with granuloma formation in the left eye (figure 1A). Considering the bilateral presentation, he was planned for OD lamellar keratectomy with living related limbal allograft (LrLAL) from his father. After explaining the details of the surgery, need for immunosuppression and the risks of failure, the procedure was performed in August 2000. Postoperatively, he attained a BCVA of 6/24 which was maintained for the next 2 months after which the procedure failed. He underwent a repeat LrLAL from his father with a penetrating keratoplasty in March 2001 after which he attained a BCVA of 20/60 which was maintained for 3 months. Between July 2001 and September 2004, he cumulatively underwent nine cultivated LrLAL surgical procedures from both his father and mother. He also underwent corneal transplantation procedures in both right (2) and left (1) eyes (figure 1B,C). Each of the procedures led to a transient clearing of his visual axis to give him vision in the range of 20/60 to 20/100 for a period of 1–3 months after which they failed. In October 2005, his BCVA was CF at 2 m in both eyes. He was given an option of Boston Ocular Scleral Prosthesis lenses which he tried but did not find to be useful. He maintained vision at CF 2 m for the next 3 years. In July 2008, he underwent cultivated oral mucosal epithelial transplantation in left eye. He attained a BCVA of CF 1 m at 1 month after which again the procedure failed. In July 2009, at the age of 15 years, he underwent a clear lens extraction with posterior chamber intraocular lens implantation with a Boston keratoprosthesis in the left eye. Figure 1D shows the preoperative status of the ocular surface when Boston keratoprosthesis was planned. On postoperative day 1 his BCVA was 20/50 (figure 1E). The posterior segment examination was performed for the first time which revealed a normal, attached retina with a cup:disc ratio of 0.2 and healthy neuro-retinal rim. Vision improved to 20/20 over the next 2 months. In his last visit at a follow-up of 41 months, he had a BCVA 20/20 with stable optic disc and visual field (figure 1F). The left eye has light perception with accurate projection of rays and a normal posterior segment on B-Scan. This has been kept in reserve with the aim to visually rehabilitate if needed in future.

DISCUSSION

This case represents a typical scenario of limbal stem cell deficiency (LSCD) in India occurring

To cite: Jain R, Sangwan VS. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-009508

Figure 1 (A) OS total limbal stem cell deficiency with granuloma formation at presentation; (B) postoperative day 1 after OS allogenic cultivated limbal stem cell transplantation; (C) postoperative day 1 after OD penetrating keratoplasty with kerato-limbal allograft in situ; (D) OS slit-lamp image showing vascularised, failed graft and failed limbal stem cell transplantation; (E) OS showing Boston keratoprosthesis in situ at postoperative day 1 and at 41 months follow-up (F).

because of exploding lime pouches resulting from the popular practice of domestic consumption of lime as an adjunct to tobacco and betel nut chewing. Most of these innocent victims are children. These injuries lead to varying degrees of LSCD, unilateral or bilateral.^{2 3}

LSCD can be treated by transplanting the limbal epithelium from a normal eye onto the diseased eye.^{4 5} Bilateral LSCD requires limbal allografting wherein the donor is a person of the same species, either living related or cadaveric.⁶ Living donors are preferred as limbal cells obtained from cadavers have a lower proliferative rate in vitro and a poorer corneal epithelisation rate in vivo.⁷ Also, the biopsy can be either used directly on the recipient or may be cultivated in laboratory. Cultivated transplantation offers significant advantages as opposed to direct transplantation in terms of reducing both the amount of donor tissue required and the corneal epithelisation time after transplantation.⁷

Our patient required 11 limbal allograft transplantation surgeries, both direct and cultivated, in a span of 5 years because of repeated failed procedures. This is in contrast to the success rate of allogenic cultivated limbal epithelial transplantation published in literature which ranges from 50% to 100% in various studies with varied follow-ups. In the study published by Basu *et al*,⁷ the technique was successful for long-term ocular surface restoration in 71.4% of the 28 recipient eyes at mean follow-up of 4.8 years. Authors hypothesise that the success of this technique could be related to the severity of the initial insult at the limbus at the time of the chemical injury. In our patient, after each surgery the ocular surface could be maintained for a transient short span of time. Though all the limbal stem cell transplantation procedures eventually failed and earlier than expected, the authors believe that a transient clear visual axis after each surgical procedure helped a great extent in the prevention of amblyopia in the paediatric age group before a final intervention was performed.

The use of Boston keratoprosthesis for visual rehabilitation in ocular surface injuries has been well documented in literature.⁸ This was performed for our patient and he now has good and

stable vision with normal optic nerve and visual fields at a follow-up of 41 months. So the authors believe that despite its various complications, Boston keratoprosthesis still is a safe and an effective long-term management option for such patients.

Management of ocular chemical injuries might be frustrating at times, both for the patient and his physician. This is because multiple ocular and extra-ocular surgeries might be required for visual and cosmetic rehabilitation. Hence, proper counselling by the treating physician and perseverance on the part of the patient and his relatives plays a very important role in treatment.

Learning points

- ▶ Ocular chemical injuries are an emergency and they need urgent intervention.
- ▶ Even if they eventually fail, repeated and timely surgeries might be required in chemical ocular injuries occurring in childhood which provide transient clear visual axis postoperatively thus preventing amblyopia.
- ▶ Perseverance is required on the part of the patient, his relatives and the treating ophthalmologist as this is a severe disease which might require multiple surgeries for visual rehabilitation.
- ▶ Boston keratoprosthesis offers an effective and a relatively safe long-term option for visual rehabilitation.

Contributors Conception and design: RJ and VSS. Acquisition of data, analysis and interpretation of data: RJ. Drafting the article or revising it critically for important intellectual content: RJ and VSS. Final approval of the version published: RJ and VSS.

Competing interests None.

Provenance and peer review Not commissioned; externally peer reviewed.

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